6-2015

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Head and Neck Pathology, Vol. 9, No. 2 (June 2015): 196-204. DOI.

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Abstract

The desmoplastic fibroma (DF) is a rare, fibroblastic lesion of bone that histologically resembles the desmoid tumor of soft tissue. Although classified as benign, it frequently demonstrates aggressive behavior, often causing tooth mobility, extensive bone destruction, and has a moderate to high recurrence rate. We present three cases of DF in the mandible: the first in a 13 year old female involving the mandibular body in the region of teeth #s 27–#28, the second in a 57 year old female with a lesion apical to tooth #30, and the third in a 20-year-old female involving the left posterior mandible. Clinical, histologic, immunohistochemical (IHC) and radiographic features of this rare neoplasm are discussed. The challenges encountered in establishing an accurate diagnosis due to significant microscopic overlap with other spindle cell lesions are also detailed. Additionally, the findings of IHC stains including vimentin, smooth muscle actin, S-100 protein, β-catenin, HHF-35 and proliferation marker, Ki-67 on 3 cases are reported. The potential for misdiagnosis is high, especially in early lesions, since immunohistochemistry has been reported in literature to be inconsistent when differentiating DFs from other spindle cell lesions. A comparative review of DF and similar entities in the jaws with current considerations in treatment and prognosis is presented.

Keywords

Desmoplastic fibroma, Desmoid tumor, Mandible, β-Catenin

Introduction

The desmoplastic fibroma (DF) of bone is considered a benign yet locally aggressive tumor of fibroblastic origin. In addition, it is also considered to represent the osseous counterpart of soft tissue aggressive fibromatosis. The clinical relevance of the DF of jaw bones lies in the controversy over its surgical management and the high rate of recurrence. Moreover, a small number of cases of DF of the jaws have been diagnosed in patients with tuberous sclerosis. Most commonly diagnosed in patients below the age of 30, DF has a predilection for the mandible, femur, pelvis, radius and tibia. When present in the jaw bones, approximately 86 % occur in the mandible and 14 % in the maxilla, with a female predilection (56 %). We describe three cases of DF in the mandible and discuss histologic characteristics, current considerations for immunohistochemical (IHC) analysis, and differential diagnoses.

Case Report #1

A 13-year-old African American female presented to an oral and maxillofacial surgeon for evaluation and treatment of an incidental right mandibular body lesion found during a routine dental visit. Clinically, teeth #27 and #28 were displaced with buccal expansion evident. Radiographically, a 2 cm multilocular radiolucency was seen between the divergent apices of teeth #s 27–28 (Fig. 1). An incisional biopsy was performed and diagnosed as a DF of the right mandible (Fig. 2a). Marginal resection of the right mandible along with extraction of teeth # 25–29 was performed. The resection sample including the three extracted teeth measured 2.5 cm (anterior–posterior) × 1.7 cm (width) × 3.0 cm (superior–inferior). Histopathologic examination of the decalcified specimen revealed trabeculae of dense viable bone surrounded by large areas of dense and sclerotic appearing fibrous connective tissue (Fig. 2a). The bony trabeculae displayed osteocytes within lacunae and no

osteoclastic activity was identified. The connective tissue was acellular and highly collagenized exhibiting band-like interconnecting fascicles of collagen bundles with minimal vascularity (Fig. 2c). Multiple IHC stains were performed, and positive labeling was noted with smooth muscle actin, vimentin, and HHF-35/muscle specific actin. Negative reactivity was seen with β-catenin and S-100, and Ki-67 <1 %.

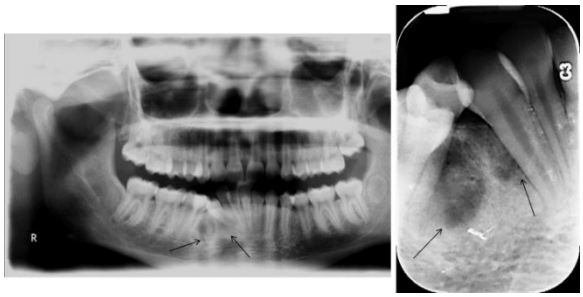

Fig. 1 Case 1. Panoramic radiograph at initial visit demonstrating the multilocular mixed lucency between apices of teeth #27–#28 and the periapical radiograph of the same area

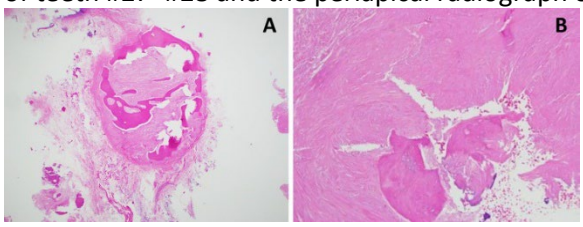

Fig. 2 Case 1. **a** Photomicrograph from the incisional biopsy sample demonstrating a spindle cell neoplasm with viable bone inside and at the periphery (H&E ×2 magnification). **b** Photomicrograph depicting dense and acellular fibrous connective tissue with viable bone, relatively acellular stroma with minimal vascularity and viable bone exhibiting osteocytes within lacunae (H&E ×10 magnification)

Based on the histologic, radiographic, and clinical features, a diagnosis of DF was rendered. The patient continues to be monitored for the last 3.5 years with no recurrence.

Case Report #2

A 57 year old Caucasian female presented to an oral and maxillofacial surgeon for implant evaluation of periodontally involved teeth #23–#26. The panoramic radiograph revealed a right mandibular 12 mm × 8 mm moderately-well circumscribed unilocular radiolucency, apical to teeth #30–#31 (Fig. 3). There was no expansion or depression of the cortical plates and the patient was completely asymptomatic. An excisional biopsy of the lesion along with extraction of periodontally involved anterior mandibular teeth #23–#26 was performed. The lesion was tenaciously adherent to the bone and displayed a soft, rubbery texture. Microscopic examination of the decalcified specimen revealed a benign fibrous proliferation composed of aggregates of dense fibrous connective tissue displaying areas of both low to moderate cellularity (Fig. 4a). The specimen was primarily composed of dense, “ropey” collagen bundles interspersed by wavy fibroblasts. The connective tissue proliferation involved regional bone and resorb bone in many foci. Trabeculae of dense viable bone appeared to be embedded within the fibrous connective tissue stroma. Foci of increased cellularity were seen within the specimen, where interlacing fascicles of collagen were noted, admixed with scattered inflammatory cells and blood vessels (Fig. 4b). IHC staining was performed, with tumor cells exhibiting

strong reactivity with vimentin and SMA focally and negative labeling for β-catenin, S-100, HHF-35/MSA, and a Ki-67 of <1 %.

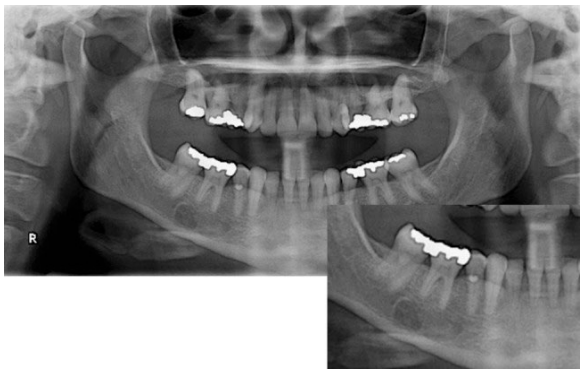

Fig. 3 Case 2. A pre-operative panoramic radiograph depicting a well-circumscribed mixed radiolucent lesion apical to teeth #s 30 and 31

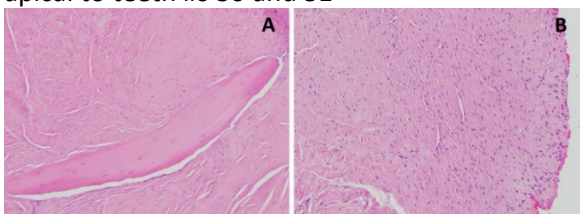

Fig. 4 Case 2. **a** Photomicrograph of the excisional biopsy depicting a fibrous proliferation with moderate cellularity with viable bone associated with the connective tissue stroma (H&E ×20 magnification). **b** Photomicrograph demonstrating bland fibroblasts with no cellular pleomorphism and no mitoses (H&E ×10 magnification)

A diagnosis of DF was rendered. She has been closely monitored over the last 3 years with no evidence of recurrence.

Case Report #3

A 20-year-old female presented for evaluation of swelling and tenderness of the left posterior mandible of unknown duration. No additional history was available. A panoramic radiograph revealed a large, multilocular, mixed radiolucent and radiopaque lesion with ill-defined borders involving the left posterior mandible (Fig. 5). Cortical thinning and expansion was noted with displacement of the mandibular canal. Tooth #17 was displaced anteriorly and inferiorly. An excisional biopsy was performed, and histopathologic examination of the decalcified specimen revealed a fibroblastic tumor composed of bland fascicles of wavy spindled cells blending with a collagenous background stroma (Fig. 6a, b). The spindled cells demonstrated moderate to high cellularity with elongated nuclei and indistinct cytoplasmic membranes. No nuclear hyperchromatism was observed. Positive IHC labeling with β-catenin and vimentin was noted in the tumor cells as well as focal positivity with SMA, and negativity for S-100 and HHF-35. The Ki-67 demonstrated <3 % positivity in the tumor cells. A final diagnosis of DF was rendered. No post-operative information was available since the patient was lost to follow-up.

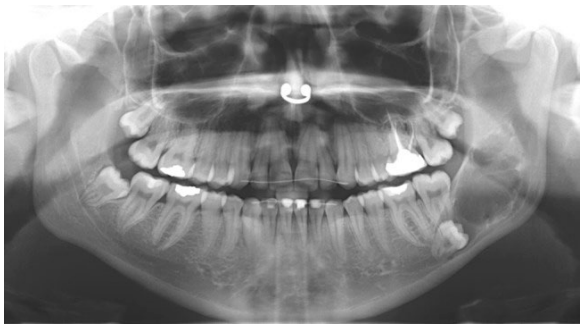

Fig. 5 Case 3. A pre-operative panoramic radiograph showing the multi-locular mixed lucent defect in the left mandibular angle and displacement of the mandibular canal and tooth #17

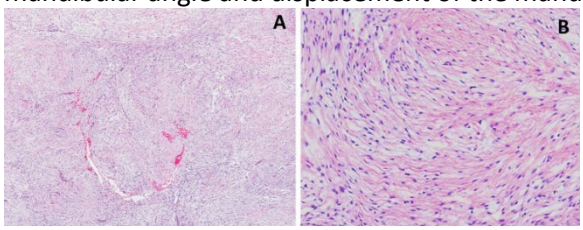

Fig. 6 Case 3. **a** Photomicrograph of a cellular neoplastic proliferation of fibroblasts in a vascular stroma. (H&E ×20 magnification). **b** Photomicrograph showing monomorphic, wavy, spindled cells with elongated nuclei in a background of collagenous stroma. (H&E ×20 magnification)

Discussion

In 1958, Jaffe introduced the term “DF of bone” to describe a densely fibrous entity composed of fibroblasts amidst rich collagen fibers, resembling the familiar abdominal desmoid tumor[1]. Griffith and Irby reported in 1965 the first gnathic involvement of a DF involving the mandible[2]. According to the World Health Organization, its low to variable cellularity, ovoid to elongated nuclei, and lack of pleomorphism and mitoses categorize it as a benign tumor [3]. DF can involve any bone but is most often found in the mandible (22 %), followed by the femur (15 %), pelvic bones (13 %), radius (12 %), and tibia (9 %)[3, 4].The incidence of DF is estimated to be 0.1 % of all primary bone tumors[3]. Approximately 84 % of patients with DF of the gnathic bones are under 30 years of age with a mean age of 16 years. Gnathically, the majority of cases involve the mandible (84 %), and in either jaw bone, a posterior location is favored. Previous statistics in 2006 from Said et al.[4] reported a slight female predilection exists among the jaw tumors (54:45 % = F:M).

While exact etiologic factors for the DF currently remain unknown, possible association with trauma, endocrine factors, genetic aberrations, or multifactorial etiology have been suggested[4, 5]. Mutations in both β-catenin and APC (adenomatous polyposis coli) genes have been implicated in the pathogenesis of desmoid-type fibromatosis[6]. Fluoroscent in situ hybridization (FISH) analyses have demonstrated trisomy 8 and 20 as nonrandom aberrations in benign fibrous conditions of both bone and soft tissue[7]. Moreover, DFs of gnathic bones have been diagnosed in patients with the hamartoneoplastic syndrome—tuberous sclerosis, in which they are believed to represent intraoral manifestations of this multisystem genetic disorder[8, 9]. Although many believe the DF to be the bony counterpart of soft tissue desmoid-type fibromatosis, others have suggested it may be a separate entity, as there have been recent discrepancies with β-catenin staining for DFs[10].

In our detailed review of gnathic DF in the English language literature[4, 9, 10, 13, 17–21, 23–52], we identified a total of 152 cases. A definite female predilection of 56 % (62/110) was noted. All three of our patients were female. Sixty-seven out of the 152 reports included ethnic information, and most cases were seen in Caucasians (45 % or 30/67), followed by African Americans (21 % or 14/67), Hispanic (12 % or 8/67), and <11 % for Asians (7/67), Middle Eastern (5/67), and South Asian/Indian (4/67). An age range of 6 months to 60 years of age, with a median age of 17 years was observed. Approximately 97 % of the mandibular neoplasms occurred in the posterior aspect, which was seen in all three of our cases. These lesions ranged in duration from 2 days to 8 years, with an average of 11.8 months. Most gnathic lesions are painless and slow-growing and the most common symptom was asymptomatic swelling (72 %), followed by facial asymmetry (20 %), tooth displacement and/or root divergence (12 %), history of previous trauma to the affected area (13 %), pain (10 %) limited mandibular opening or trismus (7 %), mimicking odontogenic infection (6 %) and tooth mobility (5 %). However, 6.3 % of patients did present with a rapidly enlarging mass. Other presenting symptoms included temporomandibular dysfunction, malocclusion, delayed tooth eruption, spontaneous bleeding, paresthesia, and pathologic fracture. Some lesions in the maxilla resulted in proptosis, palatal swelling, nasal obstruction, and sinusitis. Two of three of our patients presented with asymptomatic swelling of the jaw. One of our cases presented with expansion and tenderness. Radiographic features were reported for 56 % (84/152) cases of which 58 % presented with ill-defined borders as opposed to 42 % that had well-defined borders. Three quarters of the cases were multilocular, possibly due to the presence of thin, bony septae within the lesion, while 25 % displayed a unilocular appearance. Cortical expansion or destruction was observed in 74 % of the cases. Radiographic features of the DF affecting the jaws are not unique and overlap with many other lesions. Most DFs present with radiolucencies of ill-defined borders and non-sclerotic margins. Cortical expansion and cortical thinning are often noted, with aggressive tumors producing cortical erosion and perforation. Computed tomography (CT) or magnetic resonance imaging (MRI) is preferred over routine X-ray imaging to assess cortical continuity. Occasionally, rapid expansion following cortical perforation may result in initiation of new bone formation from the cambium or osteogenic layer of the periosteum producing a ‘sun-burst’ appearance, mimicking an osteogenic sarcoma[11]. Tumor expansion often displaces teeth and may cause root resorption. On MRIs, the majority of reported cases are osteolytic with hypointense to isointense signals on both T1 and T2-weighted images and prominent T2 shortening[12]. Wippold et al.[13] reported a hypointense lesion on unenhanced T1-weighted images that became greatly enhanced after administration of intravenous contrast medium and was hyperintense on the long TR sequences.

Treatment modalities and outcomes were documented in 72 % of the cases (109/152). The most common treatment was en bloc resection (58 %, 63/109), followed by excision/enucleation (40 %, 44/109), and curettage alone (12 %, 13/109). Chemotherapy agents, such as vincristine, doxorubicin (Adriamycin), and dacarbazine (DTIC) were used in 7 % of cases (8/109).

Follow up information was available for 120 out of 152 cases. Most cases (73 %, 88/120) were followed for time periods ranging from 3 months to 17 years, with an average of 3.3 years. No evidence of disease or recurrence was noted in 58 % (69/120), while 15 % (18/120) had documented evidence of recurrence.

Importantly, to date, no specific, reliable IHC marker(s) for labeling lesional cells in DF have been determined; therefore, DF remains a diagnosis of exclusion. This lack of markers may be explained by the possibility that acid-based chemicals used in the decalcification process may damage the tissue and interfere with binding of specific IHC agents. Since 1991, multiple IHC markers, including S100, SMA, vimentin, MSA, Ki-67, and more recently β-catenin, have been utilized to better delineate gnathic DFs from other similar entities. (Table 1) S-100 is negative in 93 % of lesions (13/14) and 63 % are negative for MSA or HHF-35 (5/8). Positive immunoreactivity is seen for vimentin in 92 % 11/12), β-catenin 50 % (4/8), SMA 77 % (10/13), and 100 % (7/7) of the lesions display <5 % Ki-67 labeling indicating a very low proliferation index.

Table 1 Immunohistochemistry in recent literature

Reference	S100	β-Catenin	Vimentin	SMA	MSA	Ki-67	Other
Boon[53]	−		+				
Hopkins[54]	unk						
Bakaeen[55]	−		−	−			
Kaplan[56]	−		+	−	−		Desmin: − CKAE1/AE3: −
Gonzalez et al.[23]	−		+	−			
Hauben et al.[10]		− +		+ +	+ +		Desmin: +, + Cyclin D-1: +, −
Said-Al-Naief et al.[4]	−		+	+f	−	<3 %	
Iatrou et al.[24]	+		+				EMA: − CKAE1/AE3: −
Dhaif[25]	−		+	+f	−		
Schneider et al.[26]	−			+		<5 %	
Mir-Mari et al.[27]	−			+			
Azola et al.[17]		+					
Taher et al.[18]		+					Desmin: + NFP: +
Gondak et al.[19]	−	−	+			<1 %	Masson's trichrome: + Picrosirus red: + CD34: − Calponin: −
Joshua et al.[28]	−		+	+f		<3 %	
Our cases							
1	−	−	+	+	+f	<1 %	
2	−	−	+	+f	−	<1 %	
3	−	+	+	+f	−	<3 %	
IHC totals:	13/14 −	4/8 +	11/12 +	10/13 +	5/8−	7/7 < 5 %	
	93 % −	50 % +	92 % +	77 % +	63 % −	100 % < 5 %	

SMA Smooth Muscle Actin, *MSA* Muscle Specific Actin or HHF-35, *EMA* Epithelial Membrane Antigen, *NFP* Neurofilament Protein, *f* focal, *unk* unknown

Interestingly, negative labeling to β-catenin was noted in of two out of three of our cases. β-catenin is a cytoplasmic protein normally located immediately beneath the cell membrane, controlled by the APC gene. It is involved in two distinct pathways involving tumorgenesis[14]. First, β-catenin binds to E-cadherin, another membrane protein, at the cell surface to form the E-cadherin-catenin complex. This

complex forms intercellular junctions, which help inhibit and suppress invasion of tumor cells. The second role of β-catenin is to mediate the Wnt signaling pathway in the nucleus to regulate cell proliferation and differentiation. This process is critical for numerous cellular processes, such as normal cell renewal and regeneration, regulation of tissue homeostasis and differentiation, maintaining normal cellular architecture, and controlling cell-to-cell adhesion[14]. Mutations in the APC gene cause an increase in β-catenin expression, which activates oncogenes and leads to a variety of neoplasms and cancers[6, 15].

Since 2002, β-catenin has been reported to positively stain neoplastic nuclei in mesenteric fibromatosis, in almost 100 % of desmoid-type fibromatoses and deep fibromatoses as well as many adenocarcinomas[6, 16]. In 2005, Hauben et al.[10] reported two cases of mandibular DFs that stained positively with β-catenin; however only one demonstrated nuclear positivity and thereby deemed true positive labeling. Multiple cases of gnathic DF have since been shown to be reactive to β-catenin[10, 17, 18]. Most recently, Gondak et al.[19] reported a maxillary lesion with negative reactivity to β-catenin (Table 1).

DFs may present with many different histologic patterns, ranging from low to moderate or high cellularity and may or may not involve viable bone. It can therefore mimic many benign and malignant entities. Histological similarities with nodular fasciitis, fibrous dysplasia (FD), myofibromas, non-ossifying fibromas, and odontogenic fibromas are noted. Of most importance is the distinction and separation of DF from FD histologically. Histopathological features of FD consist of a hypocellular fibrous connective tissue stroma with numerous irregularly-shaped, curvilinear trabeculae of woven bone, giving its “ginger root” appearance. Retraction artifact of bony trabeculae from the stroma is prominent, and there is usually no osteoblastic rimming of bone. No atypia, cellular pleomorphism, nor mitotic figures are observed in FD. Similarly, DFs are often hypocellular and share many other features with FD, making the distinction between the two extremely difficult.

Among malignant entities, the DF may histologically overlap with low grade fibrosarcoma and low grade osteosarcoma. Histopathologically, low grade fibrosarcomas demonstrate a low to moderate cellular proliferation of spindled cells, often quite uniform and without the classic “herring-bone” pattern of a high grade fibrosarcoma. Van Blarcom et al.[20], reported a case where a low grade fibrosarcoma was initially mis-diagnosed as DF. In contrast, the DF is typically hypocellular, has a more unidirectional pattern of spindle cells and nuclei with no atypia, pleomorphism, or mitotic figures. Moreover, the lesional cells in DFs display indistinct cell borders, and the stroma often encases thin spicules of trabecular bone exhibiting reactive changes.

Low grade osteosarcomas display a fibrous tissue stroma with variable cellularity consisting of spindle cells demonstrating atypia, mild pleomorphism and few to rare mitoses[30]. The connective tissue stroma can range from hypocellular to moderately cellular and contain plumper, more atypical spindled cells that resemble fibrosarcoma. The bony trabeculae are irregular, scattered haphazardly, and may have osteoblastic rimming[21]. In some cases, bone may have a “Chinese character” appearance, mimicking FD. Recent IHC studies have demonstrated positivity with MDM2 and/or CDK4 in low grade osteosarcomas, which is not appreciated in benign fibro-osseous lesions, and a Ki-67 index range of 10–37 %, which is much higher than that of the DF[22].

The comparative clinical, radiographic and histologic features and IHC findings of DF, FD and low grade fibrosarcoma are summarized in Table 2. It is important to note that recognition of these features is best accomplished when a generous sample of tissue is provided. Recognition of cellular fascicles is critical for differentiation of patterns seen in intraosseous tumors such as DF, odontogenic fibromas, myofibromas and low-grade fibrosarcomas. The biopsy should attempt to harvest tissue from the core of the lesion. A superficial biopsy involving the periphery of the tumor may show new bone formation resulting in an erroneous impression of a fibro-osseous lesion or a low grade osteosarcoma. As the DF is locally infiltrative and lacks a capsule, identification of tumor margins may be inaccurate.

Table 2 Comparison between desmoplastic fibroma, fibrous dysplasia and low grade fibrosarcoma

	Desmoplastic fibroma	**Fibrous dysplasia**	**Fibrosarcoma, low-grade**
Clinical	Asymptomatic or swelling ± pain	Slow growth with painless swelling	Slow-growing mass, ± pain
Radiographic features	Well-defined borders	Ill-defined borders	Well to ill-defined borders
	Internal pseudo-trabeculation	"Ground glass" opacification	Radiolucency with often "moth-eaten" pattern of bone destruction
	Cortical thinning with soft tissue extension	Thin cortex; lucent to sclerotic	Cortex thinned or disrupted
	May see displacement of adjacent soft tissue or bony structures	May see displacement of adjacent soft tissue or bony structures	May see displacement of adjacent soft tissue or bony structures
	MRI: osteolytic lesion with prominent T2 shortening; hypointense or isointense signal on T1 and T2 signal intensities	MRI: Not useful. Muscle like intensity on T1. Heterogenously hyperintense on T2 compared to subcutaneous fat	MRI: iso-to hypointense T1; Hypointense T2; ill-defined borders with soft tissue extension
	CT: expansion of bone with cortical destruction	CT: expansion of bone with intact overlying bone and endosteal scalloping	CT: non-specific, low density mass, with or without mixed hypodense and isodense components
Microscopic features	Thick collagenized or hyalinized connective tissue	Hypocellular connective tissue stroma	Collagenization variable; may show focal chondro-osseous differentiation or myxoid changes
	Low to moderate cellular proliferation of spindled cells	Low to moderate cellularity with bland fibroblastic cells	Low to moderate cellular proliferation of spindled cells; "herring-bone" appearance not apparent
	Thin spicules of trabecular bone with reactive changes	Numerous irregularly- shaped, curvilinear trabeculae of woven bone; "ginger root" appearance. Usually abrupt transition of normal to abnormal bone; retraction of bone from adjacent connective tissue	Calcification minimal or absent
	Uniform nuclei with no atypia, pleomorphism, or mitoses. Indistinct cell borders with cytoplasm merging with collagenous connective tissue	Uniform fibroblasts with absence of mitoses and atypia	Variable size and shape of cells: round to tapering, ovoid nuclei with increased granular chromatin, scant cytoplasm; minimal pleomorphism, few mitoses
Immunohistochemistry	Low Ki-67 Positive for vimentin, SMA, variable β-catenin Negative for S100, MSA	None	High Ki-67 Positive for vimentin, p53, Reticulin; variable CD34 Negative for S100, keratin, SMA, histiocytic markers

Treatment and prognosis

Cortical perforation and soft tissue involvement require wider margins of resection. Recurrence rates following excision and enucleation have ranged from 20 to 40 % and curettage as high as 70 %[4]. It has been noted that tumors with higher cellularity tend to recur more often than those with lower cellularity. Some authors have recommended wider resection in those tumors showing aggressive behavior as demonstrated by cortical perforation, soft tissue involvement, and moderate to high cellularity[4]. Following surgical treatment, a follow-up period of no <3 years is recommended[4].

Conclusions

The DF is a rare, benign neoplasm that can occur in gnathic bones and has overlapping histopathologic features with other benign and malignant entities characterized by spindle cell proliferations, trabeculae of bone and little to no cytological atypia or mitotic activity. Distinguishing DFs from FD, low grade fibrosarcoma, and low grade osteosarcoma has important prognostic significance and remains a difficult process for the pathologist. Ample tissue submission for diagnosis obtained from the center of the lesion is important in making a diagnosis of DF. As DFs may be locally aggressive and recur if inadequately or conservatively excised, wide resection or en bloc resection with long-term follow-up continues to be the treatment of choice based on our review. Although, some recent reports suggest the potential value of β-catenin as an IHC marker for DF, our experience (1 of 3 stained positive) did not support this claim.

Acknowledgments

We would like to thank Dr. Barton Blumberg of The Villages, Florida for his contribution to Case 2.

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