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Case Report

Recurrent Juvenile Ossifying Fibroma with Secondary Aneurysmal Bone Cyst: A Case Report

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Abstract

Juvenile Trabecular Ossifying Fibroma (JTrOF) is a rare lesion affecting mandible with a tendency toward locally aggressive behavior and recurrence. Only a few cases of JTrOF ocurring in association with secondary Aneurysmal Bone Cyst (ABC) formation have been reported in the literature. Treatment should consists of complete surgical removal because incomplete excision has been linked with a high local recurrence rate. This paper reports a case of recurrent JTrOF of the mandible with a secondary ABC in 15-year old female. The prognosis is good because malignant change and metastasis have not been reported.

Introduction

Ossifying Fibroma (OF) of the jaws is well-circumscribed, generally slow-growing lesion that enlarges in an expansile manner. The tumor is defined as a well-demarcated, occasionally encapsulated lesion consisting of fibrous tissue containing variable amount of mineralization resembling bone and/or cementum. OFs occur mostly in patients in second to fourth decades of life [1-3] The mandible is affected more often than maxilla and other cranial and facial bones. These lesions are largely restricted to tooth-bearing areas (premolar-molar region). Posterior mandibular lesions may extend superiorly into the ascending ramus. The clinical presentation of OF is usually as an expansive and painless spherical or ovoid mass. Some lesions may grow to massive size causing esthetic and functional deformity, displacing the roots of adjacent teeth. [4] Juvenile Ossifying Fibroma (JOF) is a rare fibro-osseous neoplasm that arises within the craniofacial bones of individuals under 15 years of age. Typically, the tumor involves the maxilla, paranasal sinuses, orbital and frontoethmoid bones. Mandibular lesions have also been reported. The tumor is usually asymtomatic, achieving a large size, in some cases exhibits aggressive behavior, producing significant osseous destruction with a tendency for recurrance. The occurance of multiple ossifying fibromas in the jaws is very rare [5].

Aneurysmal Bone Cyst (ABC) is a benign, pseudocystic, osseous lesion characterized by a fibrous connective tissue stroma associated with cellular fibrous tissue, multinucleated giant cells, and large blood filled spaces with no endothelial lining. ABC is an unusual bone lesion, affecting mainly the long bones, followed by vertebrae.[6] These lesions have a 1-3% prevalence for facial bones and only 1,9% of aneurysmal bone cysts occur as jaw lesion. [2,7-10] ABC occurs more frequently in the mandible (body, angle and ramus) than in the maxilla. ABCs display very different clinical symptoms, from asymptomatic slow growth to painful, rapidly growing, destructive lesions. It affects young patients under the age of 15 with no gender predilection. ABC can occur de novo or in association with lesions like ossifying fibroma, chondroblastoma, giant cell tumor of the bone, osteoblastoma, giant cell granuloma, fibrous displasia and solitary bone cyst [10-12].

This paper reports a rare case of recurrent juvenile ossifying fibroma of the mandible associated with aneurysmal bone cyst in a 15-year old female. Few cases of JOF of the mandible associated with ABC, specialy recurrent, have been reported in the literature [10-15].

A Case Report

A 15-year old female was refered to our department with a complaint of swelling in the left mandibular and submandibular region. She had first noticed the swelling in the left mandibular first molar area 3 months previously. One month later, after simple extraction of the first molar, the swelling had gradually increased in size. The patient had a history of a right nephrectomy at age 1 due to the presence of dysplastic polycystic renal disease. There was no family history of skeletal disease. Extraoral clinical examination revealed a firm mass (70x75x40 mm) in the left mandibular body The swelling extended from left second premolar posteriorly to third molar area and caudaly 15mm under inferior border of the mandibular body. No enlarged regional lymph nodes were found on palpation and Ultrasonographic (USG) evaluation. The overlying skin was normal in appearance. Intraoral examination revealed a swelling extended from the second premolar to second molar region with medial and inferior extension. The lingual cortex was expanded with no apparent mucosal changes. On palpation, the swelling was bony hard with a smooth surface and no fluctuation was evident. The patient did not complain of pain and her mouth opening was normal without any restriction or deviation. Radiologic examination (orthopantomogram) (Figure 1) revealed a radiolucency of the left mandibular body in the region from the first premolar extending to the third molar. Computer Tomography (CT) showed a large, well-demarcated mass, expanding medaily and inferiorly (Figure 2).

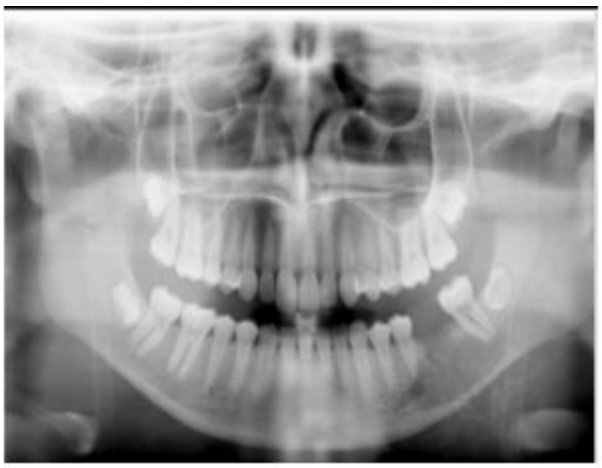


Figure 1: Orthopantomogram, radiolucent lesion of the left mandibular body.

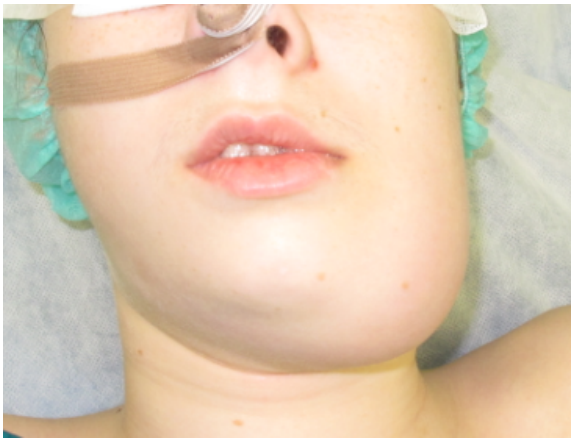


Figure 2: 3D CT reconstruction of the initial lesion.

Surgical treatment consisted of a complete resection of the left mandibular body and one stage reconstruction using a titanium 2.4 mm reconstruction plate.(Biomet, Microfixation, FL, USA) The reconstruction plate was fixed with three screws in each segment. Patient was placed on liquid diet for 6 weeks and no Maxilloamandibular Fixation (MMF) was utilized. The postoperative course was uneventful. Pathologic examination revealed a diagnosis of juvenile ossifying fibroma with secondary aneurysmal bone cyst. Clinical and radiographic follow-up 6 months postoperatively demonstrated no signs of recurrence. 10 months postoperatively, the patient developed a swelling at the symphyseal area that had rapidly expanded lingualy. Extraoral examination now revealed a midline mandibular swelling. A mandibular asymmetry was also seen below the left angle area. Intraoral examination revealed a swelling at the left lingual parasymphysis. A second expansile process was present involving the left ramus medialy and inferiorly. Both lesions had expanded mainly lingualy and medially (Figure 3). There were no mucosal changes present and the patient was pain free. CT demonstrated expansile lesions similar to the previous processes eminating from both mandibular stumps. The left ramus tumor had expanded mainly medially and inferiorly (50x60x40mm) and the medial tumor lingually (30x30x20mm) (Figures 4,5). The patient underwent complete surgical excision of the tumors consisting of a left hemi-mandibulectomy with disartculation of the condyle and resection of mandibular symphysis to the right first premolar. One stage reconstruction with a titanium 2,4 mm reconstruction was employed. Pathologic diagnosis confirmed previous diagnosis of reccurent juvenile aggressive trabecular ossifying fibroma with secondary aneurysmal bone cyst in the both mandibular stumps. Margins were free of tumor. No other bones were involved (Figure 6,7) There has been no evidence of recurence in 24 months.

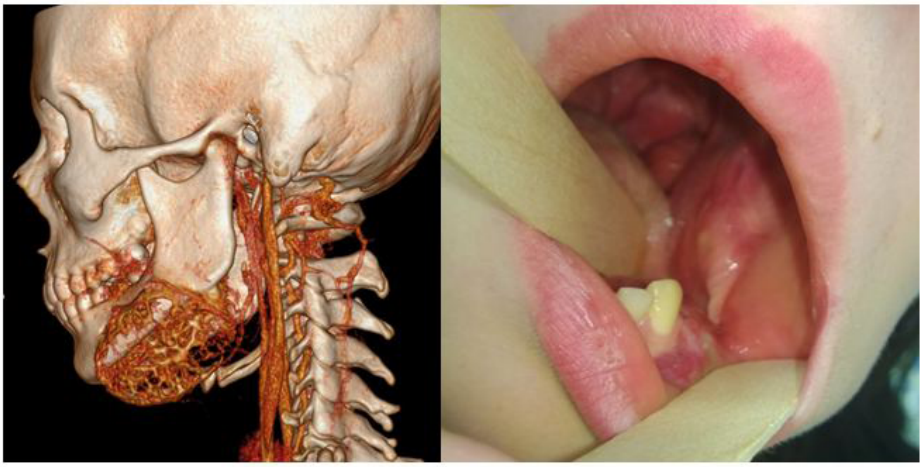


Figure 3,4: Clinical status of the reccurancy distal and symphyseal aspect.

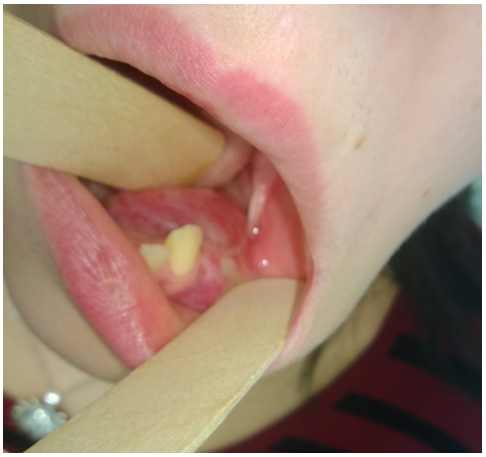


Figure 5: 3D CT reconstruction of the recurrent lesions eminating from both stumps.

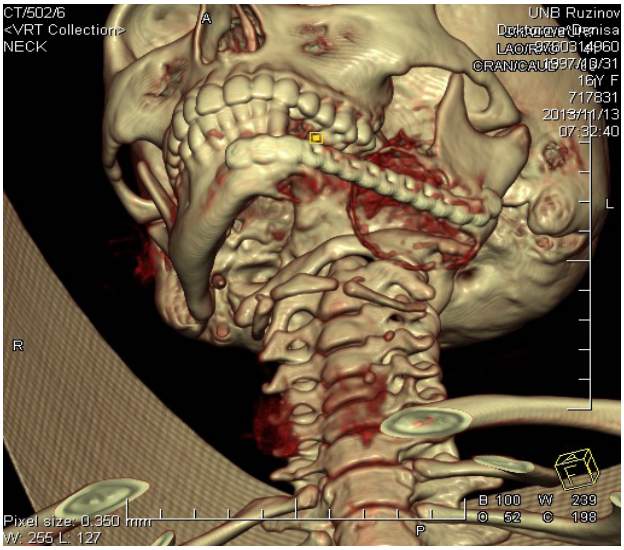


Figure 6: MRI coronal scan shows spreading of the tumor medialy and inferiorly.

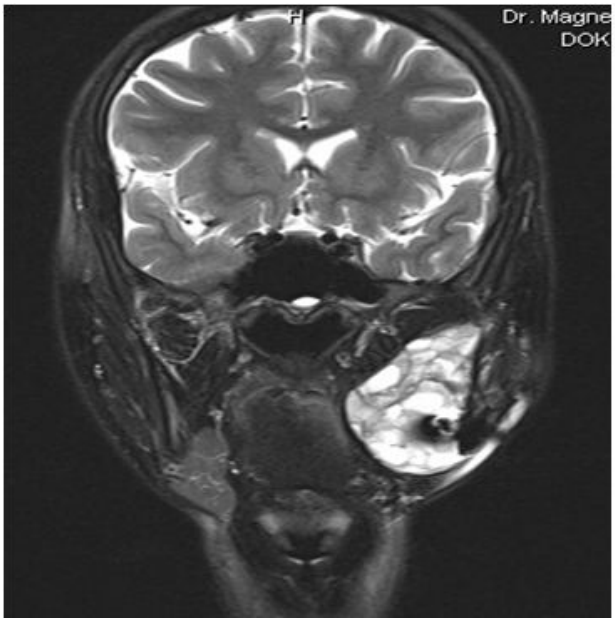


Figure 7: Surgical specimen of the mandibular left ramus.

Discussion

Ossifying Fibroma (OF) has been found in the jaws, and has also been reported in the frontal, ethmoid, sphenoid, temporal bones, orbit and anterior cranial fossa [3,16-19] Ossifying fibroma affecting the jaws exhibits a variable behavior pattern ranging from slow growth to occasionally aggressive local destruction. Although the growth of OF is described as concentric with outward expansion, approximately equal in all directions, it is unpredictable .[2,16] Juvenile Ossifying Fibroma (JOF) is also referred to as juvenile active OF or juvenile aggressive OF. The lesion is not encapsulated, although well demarcated from the surounding structures. Juvenile aggressive Ossifying Fibroma (JOF) is an uncommon, benign neoplasm of bone that presents in children and adolescents. The World Health Organisation describes JOF as a lesion affecting individuals below the age of 15. There are 2 histopathological variants of this lesion: a psammamatoid (PsJOF) and trabecular (TrJOF) type which differ entirely in their clinical and histopathologic presentation. The average age of occurance of trabecular variant is considerably younger (average age 8 – 12 years), than psammamatoid variant (average age 16 -33 years). [4,5,20] Psammamtoid juvenile ossifying fibroma (PsJOF) is a rare variant seen in the extragnathic craniofacial bones, particularly the periorbital, frontal and ethmoid bones. [4,5,15,20] Trabecular Juvenile Ossifying Fibroma (TrJOF) is predominantly a gnathic lesion affecting jaws, and there is controversy as to which jaw has a greater predilection, maxilla or mandible. There are different opinions in the literature. [2,20] The mandibular molar region is affected more often than the maxilla [17].

Both OF and JOF are almost equally distributed among both the sexes; however, JOF appears to have a male predominance. [16] Posterior segment of the jaws was primarily involved in most cases. This correlates with previous reports of cases with posterior mandible involvement.[4,7,9,21,22] Because of their slow, asymptomatic growth, they may become quite large on initial presentation , however, some lesion do demonstrate rapid enlargement, especially in younger children. The OF reccurence rate is very low. However, the recurrence rate for the JFOs vary, and have been reported as high as 30% to 56%. [2,3,16,17] This may be due to incomplete excision of the tumor. Smaller tumors may be treated successfully by enucleation and curettage. Most authors advocate resection with 5-mm margins especially for larger, aggressive tumors. Radical resection should be considered in cases where there is recurrence. [3,19,21] According to the literature, development of a concomitant Aneurysmal Bone Cyst (ABC) is more common in PsJOF than TrJOF variant. [6,11,20] The case presented here is unusual since it is associated with TrJOF. There has never been a reported case of malignant transformation of these lesions. [6,22] The term „aneurysmal“ is used to describe the expanded contour of affected bone and „bone cyst“ to indicate that the lesion appears cyctic within a thin shell of bone (Figures 8-13).

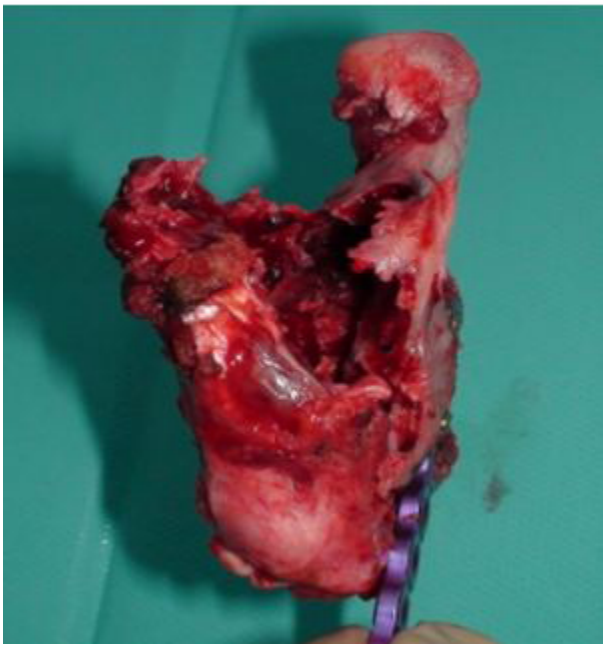


Figure 8: Clinical status 9 months after second surgery.

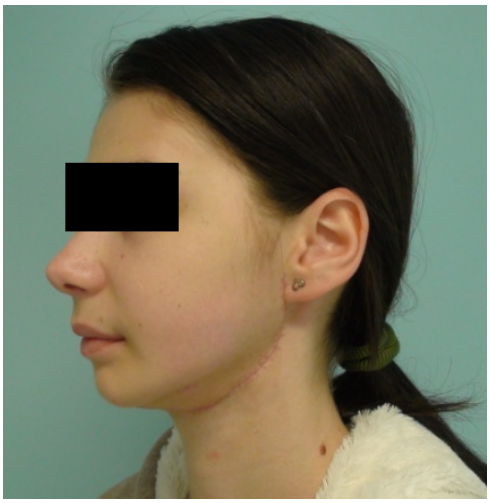
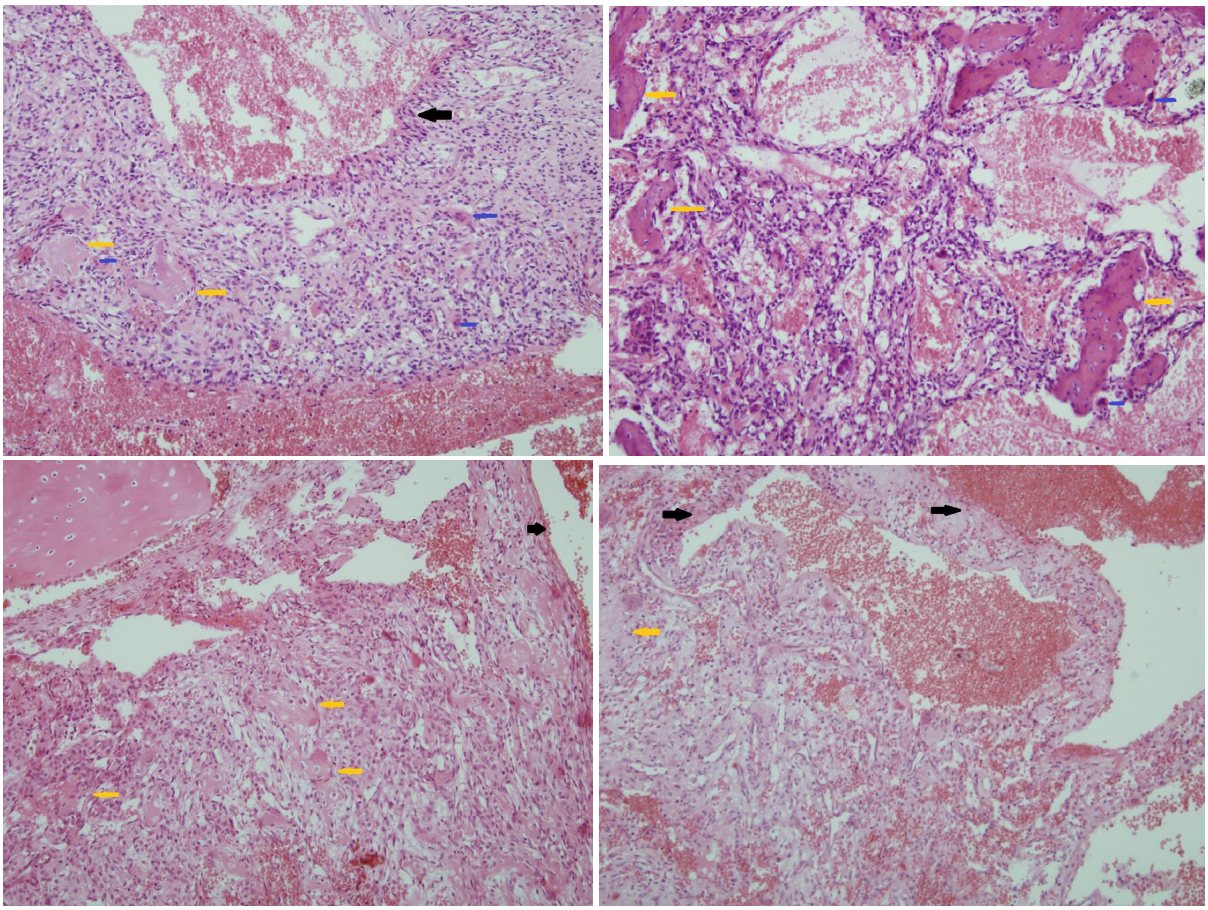


Figure 9: Present status and scar after second surgery.



Figures 10-13: Major representation of the area resembling a benign hemangioma, in addition to which structures of aneurysmal bone cyst (black arrow) are present – the cyst is most likely a secondary one, in the septums is present production of bone matrix formation (yellow arrow), including solitary osteoclasts (blue arrow). Vascularized lesion imitating a hemangioma in certain part of the histologic material, secondarily modified by aneurysmal bone cyst (black arrow), organoid trabecular deposits of bone matrix (yellow arrow), solitary osteoclasts (blue arrow).

The ABC comprises 5% of all the lesions of cranial and maxilofacial bones, although it can affect all skeletal bones, ABC primarily affects long bones followed by vertebrae but is rare in facial bones. [12,23-25] The ABC is most common in those regions of the skeleton where there is both, a relative high venous and marrow content. This partly explains the rare occurence of ABC in skull bones and jaws. [6,10,11,26,27] In the jaws, the ABC affects the mandible twice as frequently as the maxilla, and is most commonly located in mandibular body, angle and ramus. [12,27] Involvement of the mandibular condyle has been reported as extremely rare. [8,25] WHO defines the ABC as an expanding osteolytic lesion consisting of blood filled space of variable size, separated by connective tissue septa containing trabeculae of osteoid tissue and osteoclast giant cells. [2,10,28] Confusion still exist regarding the etiopathogenesis of the ABC. For a long time, the ABC was considered to be a non-neoplastic bone lesion of hemodynamic or reactive origin. This opinion was supported by elicitation of a frequent history of trauma which could induce haemorrhage within the bone. The traditional theories of ABC pathogenesis have been challenged in the last decade. The recognition of an oncogene combined with the often seen aggressive clinical presentation and high recurrence rate suggests that the ABC is not non-neoplastic lesion or pseudocyst, but should be classified as a benign neoplasm [10,23,24].

Pathologically, the ABC has been reported to exist in two forms: as a primary lesion and those secondary to other existing lesions. Histologically, the ABC can be classified as either primary or secondary, depending on the presence or absence of a preexisting lesion. Sun et al.[29] have shown that the OF is a common preexisting lesion in jaw ABCs whereas giant cell tumors are the most common in secondary ABCs involving the long bones. In the literature, various lesions were found associated with jaw ABCs. Ossifying fibroma was the most common (27%), followed by cement-ossifying fibroma (21%), central giant cell granuloma (16%), fibrous displasia (16%), cementifying fibroma (5,4%), benign osteoblastoma (5,4%), non-ossifying fibroma (2,7%), benign amaloblastoma (2,7%) and dentigerous cyst (2,7%).[10,13] There was no reported incidence of metastatic activity ABCs, or association with malignant tumours. Oncogenic evidence established that the primary ABC is indeed a neoplasm, not a pseudocyst. In contrast, the secondary ABC, due the lack of an oncogenic corelation, is actually a pseudocyst, reflecting secondary aneurysmal change in an entirely different primary neoplasm. [10,23,24] Development of an ABC secondary to other existing lesion is more common in PsJOF than TrJOF. The case presented in this report is unusual and rare since it is a case of TrJOF with secondary aneurysmal bone cyst involving the mandible. Prior published mandibular TrJOF cases do not report any ABC formation. Silva et al.[13] reported a case of TrJOF associated with ABC involving the left maxilla. Sankaranayaran et al.[14] have highlighted a recurrent case of in which an association of TrJOF was diagnosed when the lesion recurred.

No standardized treatment and follow-up protocols exisit in the literature to manage such cases. The aggressive nature of this entity with high rates of recurrence (30-50%) suggests that TrJOF with development of a secondary ABC should be treated with radical surgical resection, rather than conservative curettage. [8,12-14] The prognosis is good because malignant changes and metastases have not been reported. [10] Radiotherapy is contraindicated because of the risk of possible malignant transformation and potential harmful late effects of growth. Therefore, the appropriate recommended treatment should involve radical surgical resection of the lesion, followed by long term clinical and radiological follow-up.

Conclusion

Juvenile ossifying fibroma, associated with an aneurysmal bone cyst is locally aggressive and has a high reccurance rate when not adequately treated initially. In the case presented, despite initial radical en-bloc resection, demonstrating tumor free surgical margins, the lesion recurred. This report emphasizes that despite primary radical tumor resection and clinical follow-up, the presence of such an aggressive pathologic entity creates risk of future reccurance. Therefore, these lesions should be treated aggressively initally.

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