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CASE REPORT

Management of juvenile ossifying fibroma in the maxilla and mandible

Sondos Abuzinada[1]✉ · Alyamani A[2]

[1] Resident
[2] Associate Professor

King Abdulaziz University, Faculty of Dentistry, Saudi Arabia

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Abstract We present three cases with juvenile ossifying fibroma. Two occurring in the maxilla, and one in the mandible. All three cases presented with a major swelling in the face. After clinical and radiological evaluation the lesions were surgically excised and sent for histopathological evaluation. Two histological types of juvenile ossifying fibroma were found, the psammatous type in two cases and the trabecular pattern in one case. Although juvenile ossifying fibroma is an uncommon clinical entity, its aggressive local behaviour and high recurrence rate mean that it is important to make an early diagnosis. It is also important to apply the appropriate treatment and to follow-up the patient closely over the long term. This report describes the diagnosis and treatment of juvenile ossifying fibroma in the maxilla and the mandible. It also emphasizes the importance of considering the less aggressive options as a first line of treatment before choosing the aggressive approach when dealing with children.

Keywords Juvenile ossifying fibroma · Fibro-osseous lesions · Maxilla · Mandible · Cemento-ossifying fibroma

Address for correspondence:

Sondos Abuzinada
P.O.Box 419
Jeddah
Saudi Arabia
E-mail: sondoz@hotmail.com

Introduction

Juvenile Ossifying Fibroma (JOF) is a rare expansile fibro-osseous tumour which can arise in the craniofacial bones. The patient's age at onset distinguishes it from other fibro-osseous lesions as primarily it appears in individuals under 15 years of age.

JOF is usually asymptomatic, achieving large size and exhibiting an aggressive behaviour, and it is often diagnosed as 'juvenile ossifying fibroma,' 'aggressive ossifying fibroma,' or 'active ossifying fibroma.' The literature notes two histological types of JOF. The psammatous and the trabecular type [1].

JOF demonstrates an aggressive local behavior, and a high recurrence rate. This highlights the importance of making an early diagnosis, applying the appropriate treatment and, most importantly, following up the patient closely over the long term [2]. Most JOF arise near the paranasal sinuses [3], but there are conflicting reports regarding the incidence of JOF [2,4] in the maxilla, and mandible [5].

This report describes three clinical cases with two different types of JOF. Two cases with the psammatous type and one with the trabecular type. We discuss in detail their clinical, radiological and histological presentation. We also discuss the treatment modalities used for each case.

Cases

The first case was a 6-year-old boy with a clear medical history. He was referred to the Oal and Maxillofacial Surgery, Department at King Abdulaziz University Hospital, with a swelling in the left side of his face (Fig. 1a). It had appeared nine months earlier and had continued to increase in size. Clinical examination revealed a hard mass in the left maxillary region. It was asymptomatic with no pain nor paresthesia. The overlying skin was normal and no lymphadenopathy was noted. Intra-orally there was a clear buccal expansion in the left maxillary region, but the overlying mucosa and related teeth had normal findings. The patient had no visual disturbances and his chest x-ray was clear. The panoramic radiograph showed a well defined radiolucent lesion confined to the left maxilla. The CT showed a well defined radiolucent mass measuring approximately 3x3cm involving the left maxillary region, alveolar bone, and sinus. The orbital floor was slightly elevated which caused a displacement of the orbital contents leading to a slight vertical dystopia which was appreciated clinically (Figs. 1b and c).

The patient underwent surgical curettage of the lesion using an intra oral buccal sulcus incision and a Caldwell luc approach to the sinus, the lesion was excised with the associated developing tooth buds. The mass was fibrous in consistency and the area was curetted until a clear surgical bed was achieved. A drain was placed into the sinus extending out of the left nostril, which was removed 24 hours later. The biopsy report showed ossifying fibroma; microscopically showing 'C' shaped bone trabeculae with some osteoblast rimming and surrounding proliferating fibroblastic stroma. Blood vessels and areas of granulation tissue were also seen, while in other areas foci of cement-like material with psammoma-like bodies were found (Fig. 1d). Six months later, during follow-up, a new CT revealed bony regeneration (Fig. 2a). A year later the patient presented with a mild left facial swelling. The CT revealed mixed

radioopaque and radiolucent formation in the floor of the orbit and the lateral nasal wall (Fig. 2b). Under general anaesthesia the area was curetted and the tissues sent for histopathological evaluation which confirmed a recurrence of JOF. Since then the patient has been under regular follow-up. He was seen once in every six months for two years. He received a CT examination every six months during the first year, then once in the second year. His most recent CT showed bony regeneration in the area and did not show any bony changes in comparison to the previous CT taken a year earlier (Figs. 3a and b) with no clinical signs of recurrence (Fig. 3c). Based on the clinical observation and the radiographic assessment the patient was considered free of recurrence and continued to demonstrate the benefit of our conservative surgical treatment after two years of follow-up.

The second case was a 4-year-old female. She presented with a left facial swelling, which had become apparent five months earlier and had continued to increase in size (Fig. 4a). Upon clinical examination a large mass was discovered which extended from the left maxillary region with a severe palatal expansion. No visual disturbance was noted, but a vertical dystopia and a slight exophthalmos was clinically obvious. Her left nasal airway was completely obliterated with a clear nasal deformity. Her panoramic radiograph showed a well defined radiolucent lesion in the left maxilla. The CT showed an extensive well defined radiolucent mass measuring approximately (5x4cm) with some areas of central calcification extending from the left maxilla anteriorly into the infratemporal fossa posteriorly, involving the lateral nasal wall, leading to a septal deviation towards the right side. The orbital floor was elevated causing displacement of orbital contents (Figs. 4b and c). Due to the extensive size of the mass a left partial maxillectomy was done using a Weber – Ferguson flap approach. The orbital floor and anterior maxillary wall were reconstructed using calvarial cortical blocks, fixed using wires. The aggressive destruction and extensive size of the lesion required an aggressive surgical approach. Microscopically, sections revealed proliferation of connective tissue with plump spindle cells and numerous irregular bone trabeculae containing osteocytes in their lacunae. Some of the bone trabeculae were focally rimmed by osteoblast proliferation. Areas of bone resorption with osteoclast giant cells were also seen (Fig.

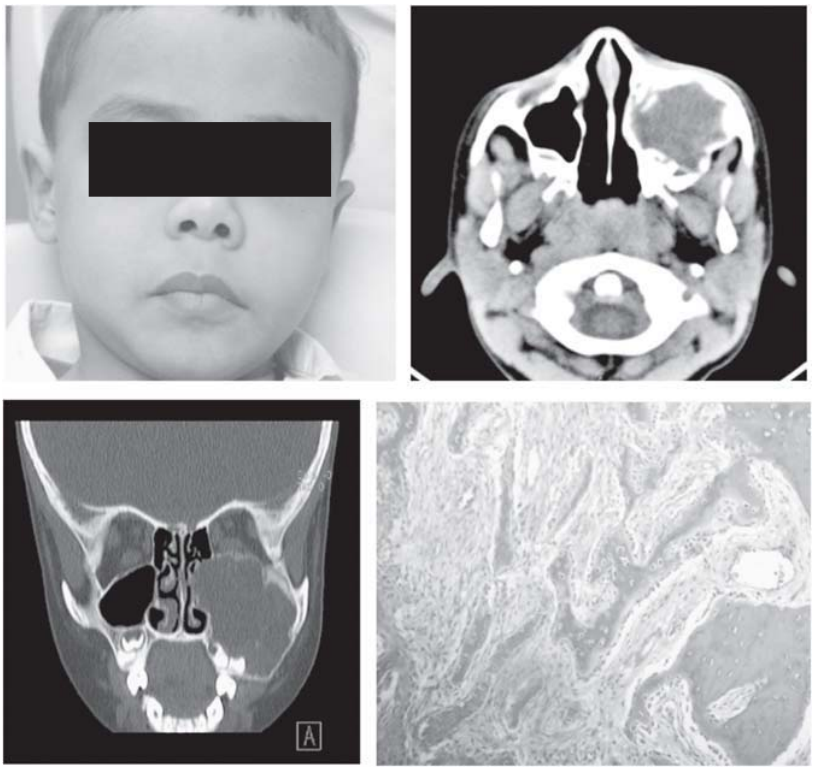

Fig. 1 (a) Facial appearance of swelling in the left side (b) Axial Computed tomography (CT) image of skull showing well defined radiolucent mass involving the left maxillary region (c) Coronal (CT) image of skull showing well defined radiolucent mass involving the left maxillary region, alveolar bone, and sinus and the orbital floor (d) Microscopic image showing bone trabeculae with osteoblast rimming and surrounding proliferating fibroblastic stroma. Blood vessels and areas of granulation tissue were also seen (H&E 20x/0.40)

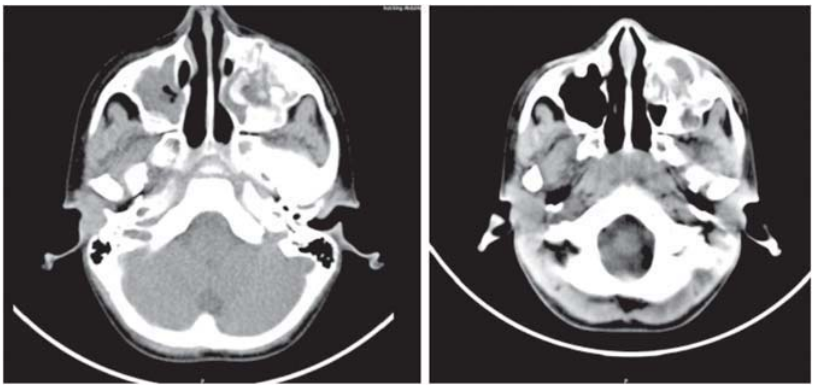

Fig. 2 (a) Axial CT showing bony regeneration 6 months post curettage with thick radio-opaque margin (b) Axial CT showing recurrence of mixed radiolucent and radio-opaque lesion a year later

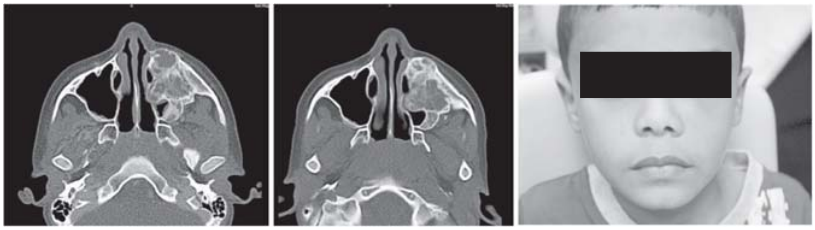

Fig. 3 a) Axial CT after one year follow-up showing mixed radiopaque and radiolucent lesion in the left maxillary region b) Axial CT after two years follow-up showing no changes c) Clinical picture after two years follow-up showing no clinical changes

4d). Accordingly, in conjunction with the clinical picture, the lesion was diagnosed as JOF. The patient was under close follow-up. She was assessed once in every 6 months for two years. She remained clinically and radiographically free of any recurrence.

The third patient was a 9-year-old boy with a painless swelling in the left side of his face. The patient and his parents had noticed that the size of the swelling had increased slowly over time. There was no history of trauma nor infection. Physical examination revealed a healthy young boy with facial asymmetry caused by an expanding mass in the left side of the mandible with no lymphadenopathy. Intra-oral examination showed expansion of the buccal cortex with no evidence of tooth mobility and no malocclusion. His panoramic radiograph showed a well circumscribed radiolucency related to unerupted mandibular second molar measuring approximately 2x2cm with some central opacity (Fig. 5a). An incisional biopsy revealed a fibro-osseous lesion favouring ossifying fibroma. The mass was curetted with peripheral ostectomy of the bony cavity and removal of the related unerupted tooth. Microscopic examination of the specimen showed spindle cells and fibroblastic proliferation with bony trabeculae, and osteoid. Some osteoblastic rimming and psammoma–like calcified masses mitotic figures were seen (Fig. 5b). Based on the clinical and histological features of the lesion, a diagnosis of JOF was reached. He was under close follow-up. He was assessed once in every three months. His OPG six months postoperatively showed bony regeneration (Fig. 6a). Eighteen months later, the same patient presented with a swelling in the same area. His panoramic radiograph showed a well defined radiolucent area in the left side of the mandible body extending to the angle and ramus measuring approximately 2x4cm (Fig. 6b). An incisional biopsy showed a recurrent fibro-osseous lesion, which correlated with recurrent juvenile ossifying fibroma. The patient underwent a mandibular block resection with placement of a reconstruction plate. The patient received very close follow-up assessments. He was examined once in every three months for two years. He was transferred to another hospital for undergoing mandibular reconstruction.

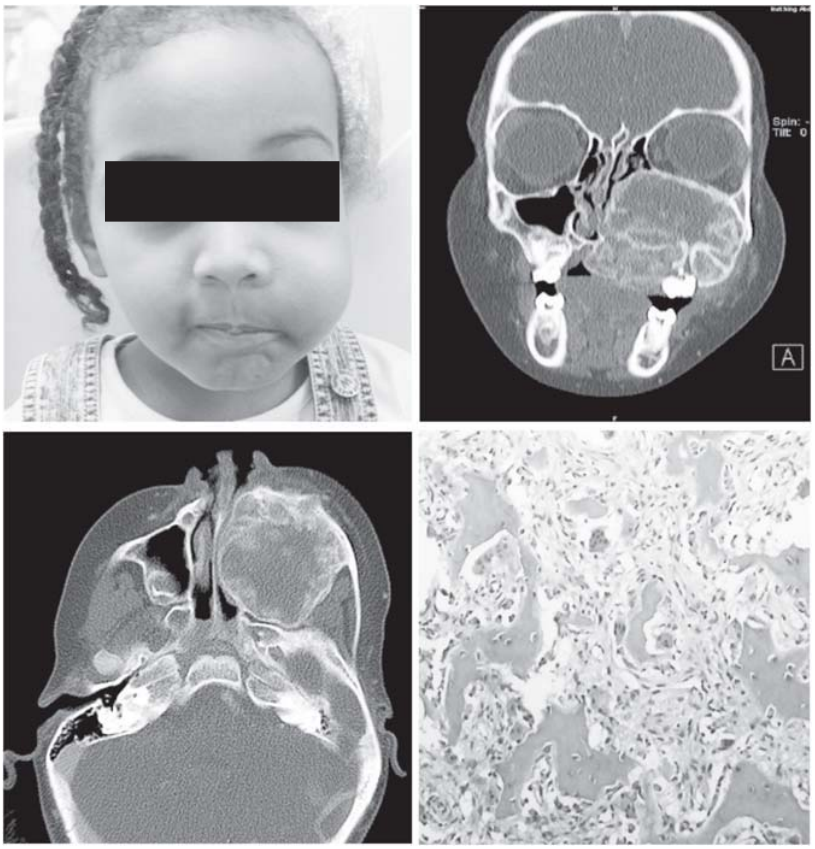

Fig. 4 (a) clinical picture showing swelling in the left side of the face (b) coronal CT showing a well defined radiolucent mass with some areas of central calcification, involving the left maxilla, maxillary sinus, nasal cavity ,alveolar bone with orbital floor displacement (c) axial CT showing the lesion invading the lateral nasal wall with septal deviation (d) Microscopic image showing proliferation of connective tissue with plump spindle cells and numerous irregular bone trabeculae containing osteocytes and some areas of bone resorption with osteoclast giant cells (H&E 40x/0.65)

Discussion

Ossifying fibroma of the jaw was first described by Montgomery in 1927, as a benign fibro-osseous lesion [5]. Differentiating between fibrous dysplasia and ossifying fibroma may be difficult. Generally, ossifying fibroma is a well circumscribed lesion unlike fibrous dysplasia and it is usually surrounded by a fibrous capsule [6]. Ossifying fibroma may exhibit a sclerotic margin on plain radiographs and CT [7].

Juvenile Ossifying Fibroma (JOF) also known as 'aggressive,' 'active,' or 'psammomatoid' ossifying fibroma is a variant of ossifying fibroma [8]. The term 'juvenile ossifying fibroma' was first used by Johnson in 1952, when he was describing aggressive forms of ossifying fibroma as it occurred in the craniofacial bones of children [9]. The literature notes the difficulty in establishing a definitive diagnostic criteria for JOF from both a clinical and a histopathological point of view. However, it is mainly characterized by occurring in patients under 15 years of age and having a high tendency to recur [2]. Clinically it may present as an asymptomatic gradual or rapid expansion of the affected bone leading to facial asymmetry. It can grow to a considerable size and show an aggressive behaviour of rapid growth with cortical thinning and perforation [10]. Radiographically, it can be seen as a unilocular or multilocular radiolucency with well defined borders; occasionally some central opacification is observed [11]. Radiographs can show root displacement and resorption though rarely [1].

CT examination may exhibit a radiolucent or mixed radiolucent and opaque lesion with well defined borders and a thin sclerotic shell. JOF has a predominantly soft tissue consistency with a variable amount of internal calcification. It shows a more invasive and destructive pattern on CT compared with conventional ossifying fibroma [12].

JOF is defined as a fibro-osseous lesion that is characterized by cell-rich fibrous tissue with bands of cellular osteoid

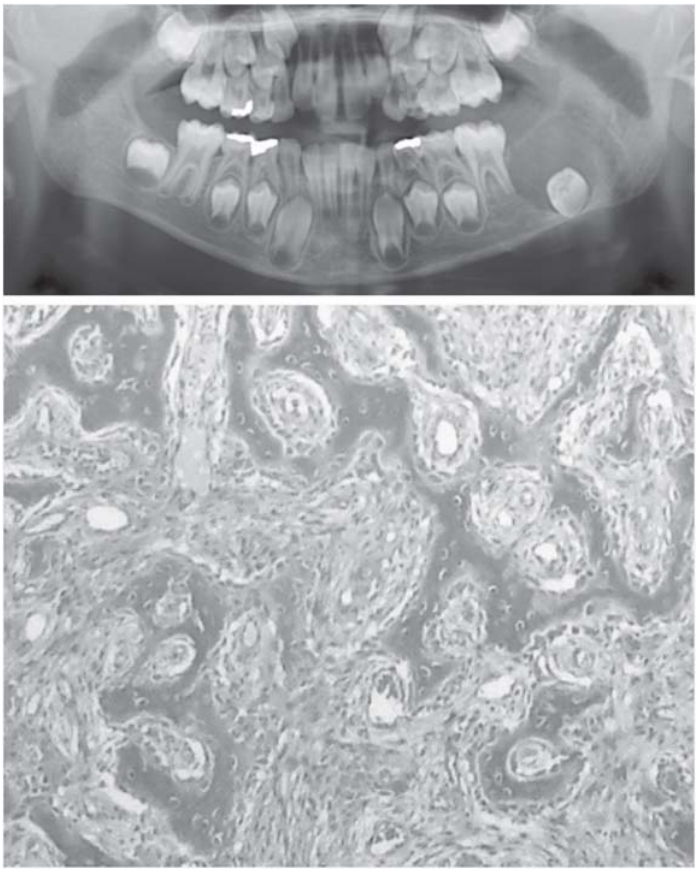

Fig. 5 (a) OPG showing a well defined radiolucent lesion in the left side of the mandible, related to the second molar (b) Microscopic image showing bone trabeculae with some osteoblast rimming and surrounding proliferating fibroblastic stroma. Blood vessels and psammoma–like calcified masses (H&E 20x/0.40)

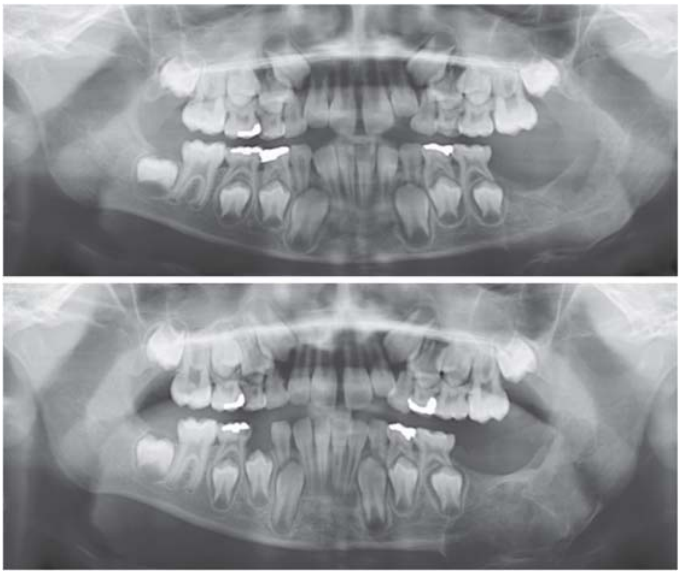

Fig. 6 (a) Postoperative OPG showing the bony defect where the mass was curetted and after removal of the related unerupted second molar (b) OPG taken 18 months postoperatively showed a well defined radiolucent area in the left side of the mandible body extending to the angle and ramus

trabeculae, and giant cells [13]. However, there is a degree of overlap between JOF and cemento-ossifying fibroma, which is histologically characterized by a uniform pattern. JOF is more likely to show markedly heterogeneous morphology. This is a characteristic that makes the diagnosis more difficult and challenging. Areas of dense cellularity may alternate with myxomatous regions, and the distribution of bone trabeculae and ossicles are often uneven. However, giant cells are clearly found in JOF, but they are not found in cemento-ossifying fibroma [14,15]. Therefore, the interpretation of incisional biopsies of these lesions must be undertaken carefully and accurately in order to establish a correct diagnosis. The rapid growth rate often exhibited by JOF can be alarming and may cause the clinician to suspect the presence of malignancy. Therefore, it is extremely important to maintain good communication between surgeon and pathologist in order to confirm the benign nature of the lesion and prevent over treatment [16]. Pain and paresthesia are rare manifestations. Nasal obstruction, exophthalmos and intracranial extension could be associated with lesions arising in the orbit, paranasal sinuses and maxilla [17].

The term JOF, as used in the literature, describes two microscopically distinct fibro-osseous lesions of the craniofacial skeleton. One is characterized by small uniform spherical ossicles resembling psammoma bodies (psammomatoid juvenile ossifying fibroma), as seen in the first and third cases. The second is distinguished by trabeculae osteoid and woven bone (trabecular juvenile ossifying fibroma). Aggressive growth occurs in some but not all cases of both types. Such behaviour may be related to younger patients and the concurrent development of aneurysmal bone cysts, which is seen more frequently in psammomatoid juvenile ossifying fibroma [1].

Congenitally missing permanent teeth have been associated with JOF. Noffke reported a case of JOF in a 4-year-old boy with a missing first molar in the same site [10]. Moreover, Johnson et al. reported a recurrent JOF in the mandible of a 12-year -old girl who was missing the permanent first molar. This association could be explained by a disturbance in the formation of the tooth socket by the JOF, which in some cases may obliterate the involved developing tooth [17].

Treatment of JOF is either conservative or aggressive according to the behaviour of the lesion. Conservative treatment of

non-aggressive forms of JOF includes curettage, as performed in the first case, to avoid violation of the orbital floor and, in the third case, due to the small size of the lesion and to preserve the inferior alveolar nerve. Conservative treatment also includes local excision as carried out in the second case to preserve the surrounding vital structures [18]. Children with recurrent JOF can be managed by local surgical excision [19]. Aggressive lesions, which show rapid growth, cortical bone thinning or perforation, tooth displacement or root resorption can exhibit early recurrence. Therefore a block resection is justified when extremely aggressive behaviour is observed [20]. However, we treated the recurrence in the first case conservatively with curettage and close follow-up, to spare the child the agony of dealing with an orbital deformity and the possibility of losing his eye. Which would have resulted from an aggressive approach. On the other hand, a conservative approach using curettage could not be applied to the third case due to the severe cortical bone thinning. Curettage would have caused a mandibular fracture and a loss of continuity in this case, therefore a block resection was the treatment of choice.

The recurrence rate for JOF ranges from 30% to 58%, which is considered high [17], consequently regular follow-up assessments are essential and their importance cannot be over-emphasized in such cases, especially if a conservative treatment was choosen.

Kaban and colleagues have suggested thorough curettage or enucleation with adjuvant interferon alpha therapy for one year when treating children with aggressive JOF in the maxilla, orbit, or paranasal sinuses. Subcutaneous interferon alpha has an antiangiogenic effect and has proven to be effective in treating giant cell lesions after enucleation or curettage [21]. To the best of our knowledge there are no published articles on the use of interferon alpha in treating JOF, so further research in that area would be of great benefit. Interferon alpha could have been used to manage the recurrence in the first case, but it was unavailable at that time.

When dealing with JOF the less aggressive approach should always be considered as the first line of treatment in order to spare children extensive facial deformation and loss of vital structures. We have demonstrated the great benefit of conservative treatment in managing JOF with an emphasis on the vital importance of vigilance and close follow-up in such cases.

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