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AN INFLAMMATORY MYOFIBROBLASTIC TUMOUR PRESENTING WITH LIMITED MOUTH-OPENING, HYPOESTHAESIA OF THE LEFT CHIN AND INFRAORBITAL AREA, INTERMITTENT LEFT EYE PTOSIS AND CONVERGING STRABISMUS

Imen Ben Hassine[1,2,3], Wiem Romdhane[1,2,3], Raouaa Belkacem[4], Jihed Anoun[1,2,3], Wafa Baya[1,2,3], Anis Mzabi[1,2,3], Fatma Ben Fredj[1,2,3]

[1] Faculty of Medicine, University of Sousse, Susa, Tunisia
[2] Internal Medicine Department, Sahloul University Hospital, Susa, Tunisia
[3] Geriatric medicine and Gerontology Unit, University of Sousse, Susa, Tunisia
[4] Department of Oral Medicine and Oral Surgery, Sahloul University Hospital, Susa, Tunisia

Corresponding author's e-mail: wiem.romdhane5@gmail.com

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ABSTRACT

An inflammatory myofibroblastic tumour (IMT) is a rare neoplasm of mesenchymal origin, defined by myofibroblastic spindle cells accompanied by inflammatory cells, lymphocytes and eosinophils. Its symptomatology depends on the involved site and tends to mimic a malignant tumour clinically and radiologically. The head and neck region accounts for 5% of all IMTs. Here, we report a case of a 35-year-old woman, with no medical history, who presented with a mouth-opening limitation of 8 mm evolving for three years and occurring six months after of a wisdom tooth extraction. She also experienced a recent occurrence of left eye ptosis and a converging strabismus. On examination, the patient had a body temperature at 37°C, with hypoesthaesia of the left chin and infraorbital area, without any other abnormality. Laboratory examinations did not reveal a biological inflammatory syndrome or rhabdomyolysis. The infectious investigations were all negative, as well as the immunological tests, in particular negative for anti-AChR and anti-MuSK antibodies. On the facial computed tomography (CT) scan, we noted an active reshuffle in the left mandible ascending branch with a thickening of the ipsilateral pterygoid muscles and the left temporal meningeal tissue. After corticosteroid therapy 0.7 mg/kg/j, we obtained an improvement in the patient's mouth-opening, thus a biopsy of the lesion was performed under local anaesthesia, revealing IMT. The patient continued the corticosteroids therapy with a progressive tapering resulting in a marked clinical improvement of the mouth-opening limitation and her ptosis.

KEYWORDS

Myofibroblast, neoplasm, ptosis, steroids

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LEARNING POINTS

- An inflammatory myofibroblastic tumour (IMT) is a challenging disease.
- Given the variable clinical and radiological presentation of the disease, it is of paramount importance to know it, to be swiftly recognised so diagnosis can be promptly made.
- The adapted treatment should be immediately started to prevent possible life-threatening outcomes.

INTRODUCTION

An inflammatory myofibroblastic tumour (IMT) is a rare neoplasm of mesenchymal origin, composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils[1]. It has been classified as a neoplastic disease of intermediate biological potential due to its low risk of recurrence and metastatic potential[2].

Previously described as inflammatory pseudotumour, IMTs are now considered as distinct tumours with specific histopathologic features. It most commonly affects children and young people with a prevalence ranging from 0.04% to 0.7%, regardless of gender and race, in the world population[3].

It usually represents both diagnostic and therapeutic challenges since the symptomatology depends on the involved site and it tends to mimic a malignant tumour clinically and radiologically[3]. IMT affects most often the lungs, abdomen, pelvis and retroperitoneum. However, it has been reported in other locations, including the head and neck region, which accounts for 5% of all IMTs and 14% to 18% of extrapulmonary localisations[3].

Here, we report a case of mandibular localisation of IMT with a particular clinical presentation as ptosis and mouth-opening limitation.

CASE DESCRIPTION

A 35-year-old woman with no medical history presented with a mouth-opening limitation of 8 mm *(Fig. 1)*. On examination, she had a body temperature of 37°C, with hypoesthaesia of the left chin and infraorbital area. There was no cough, no chest pain, no abdominal pain, no weight loss or other abnormality.

Laboratory examinations did not reveal a biological inflammatory syndrome or rhabdomyolysis. The patient presented with a mouth-opening limitation of 8 mm *(Fig. 1)* evolving for three years and occurring six months after a wisdom tooth extraction. She also experienced a recent occurrence in the previous three months of left eye ptosis and a converging strabismus without any other abnormality. The infectious investigations were all negative, as well as the immunological tests, in particular negative for anti-AChR and anti-MuSK antibodies. On the facial computed tomography (CT) scan, we noted an active reshuffle in the left mandible ascending branch with a thickening of the ipsilateral pterygoid muscles and the left temporal meningeal tissue *(Fig. 2)*. The cerebro-orbital magnetic resonance imaging (MRI) showed a clear enhancement of the pterygoid muscles and the left infratemporal fossa, extending to the ipsilateral foramen ovale, which is enlarged, with enhancements of the meninges, the cavernous sinus and Meckel's cave.

After corticosteroid therapy 0.7 mg/kg/j, there was an improvement in her mouth-opening *(Fig. 3)*, so a biopsy of the lesion was performed under local anaesthesia *(Fig. 4)*. The histopathological examination revealed the proliferation of fibroblasts and myofibroblasts, accompanied by mixed

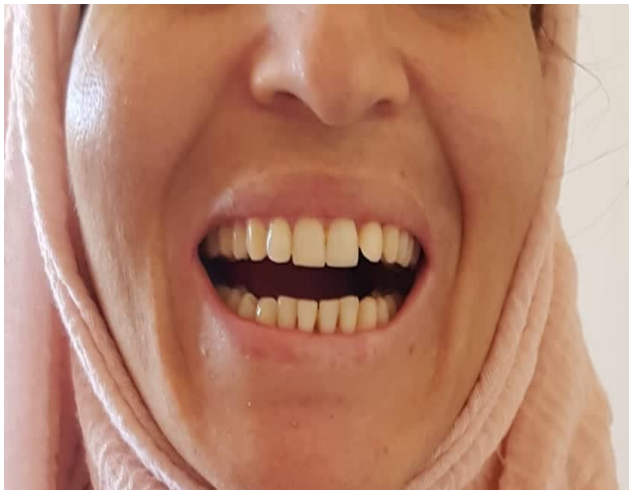

Figure 1. Limitation of month opening to 8 mm.

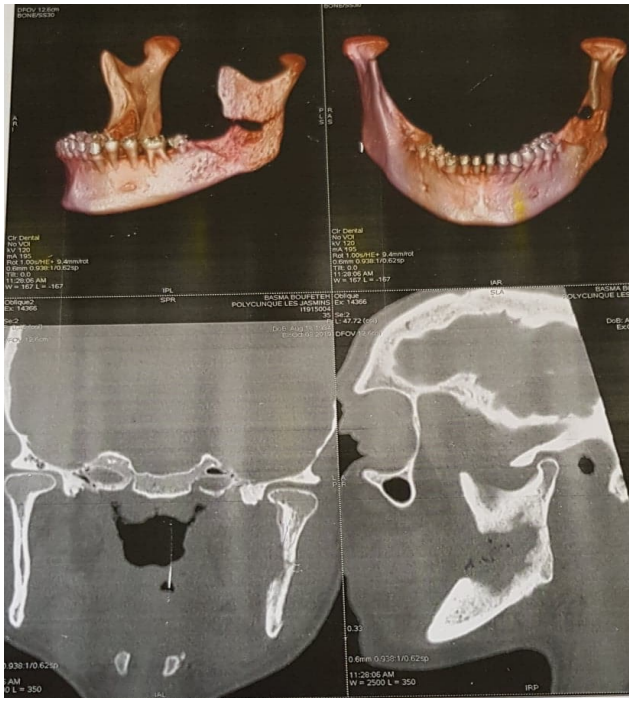

Figure 2. Reshuffle in the left mandible ascending branch.

Figure 3. An improvement in the patient's mouth-opening after the corticosteroid therapy.

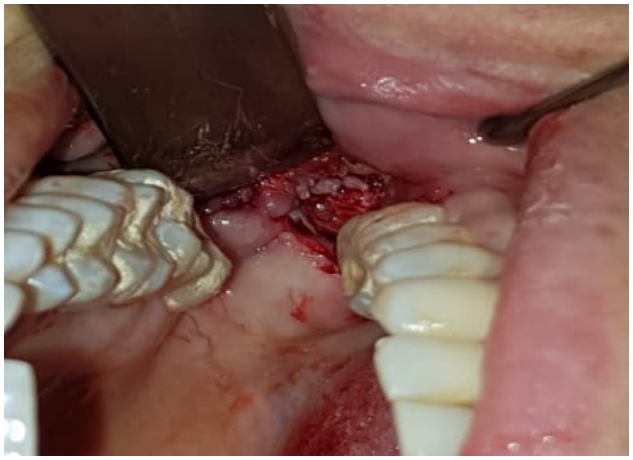

Figure 4. The biopsy of the lesion was performed under local anaesthesia.

inflammatory infiltration by plasma cells, lymphocytes, eosinophils and histiocytes compatible with IMT *(Fig. 5)*.

The patient continued the corticosteroids therapy with a progressive tapering resulting in marked clinical improvement of the mouth-opening limitation and her ptosis. She remained stable and on the follow-up imaging, the MRI orbit-face neck showed the same appearance with no signs of the disease activity and the regression of left temporal meningeal contrast *(Fig. 6)*. Hence, the patient did not undergo a complete surgical resection of the tumour due to the favourable clinical outcome and the risk of mutilations, so her corticosteroid treatment was discontinued.

The patient experienced fronto-temporal and occipital headache without associated signs, three months after the discontinuation of the oral corticosteroids. On imaging, there were no signs of disease activity; ophthalmic, and ear, nose and throat cause of headache were ruled out. We reintroduced corticosteroid therapy, and there was a regression of headache. The patient remained stable under corticosteroid therapy at a low dose (0.1 mg/kg/j).

DISCUSSION

IMTs are rare mesenchymal tumours. They can occur due to a genetic mutation characterised by the rearrangement of the anaplastic lymphoma kinase (ALK) gene on chromosome 2p23 leading to activation of the ALK of the tyrosine kinase in approximately 36–60% of all patients with IMT[3]. Other molecular alterations have also been identified in IMTs[4].

Although the aetiology of IMTs remain uncertain, numerous aetiologies may be involved in its pathogenesis such as infection, autoimmune diseases, foreign bodies and radiation[3]. Moreover, dental treatment and trauma have been suggested as linked its occurrence[3] and we speculate that in our case, those two conditions may be the cause of the IMT.

In the head and neck region, it has been found in parapharyngeal spaces, the maxillary sinus, epiglottis and oral cavity. However, intrabony IMTs are rare with only a few reported cases of IMT involving the mandible. Clinically, it is most commonly painless with an indurated mass, or it may be manifested by a swelling[5]. In our case, the tumour was localised in the mandible and then it infiltrated the adjacent structures resulting in ptosis and a mouth-opening limitation of 8 mm. To our knowledge, it is a rare localisation, and this is the first case to describe such a clinical presentation of IMT. The clinical polymorphism of IMT requires pathological proof for diagnosis and for therapeutic management.

The radiological findings of IMTs are non-specific[6]. On ultrasonography, they can appear as hypoechoic or hyperechoic masses with well-circumscribed edges[6].

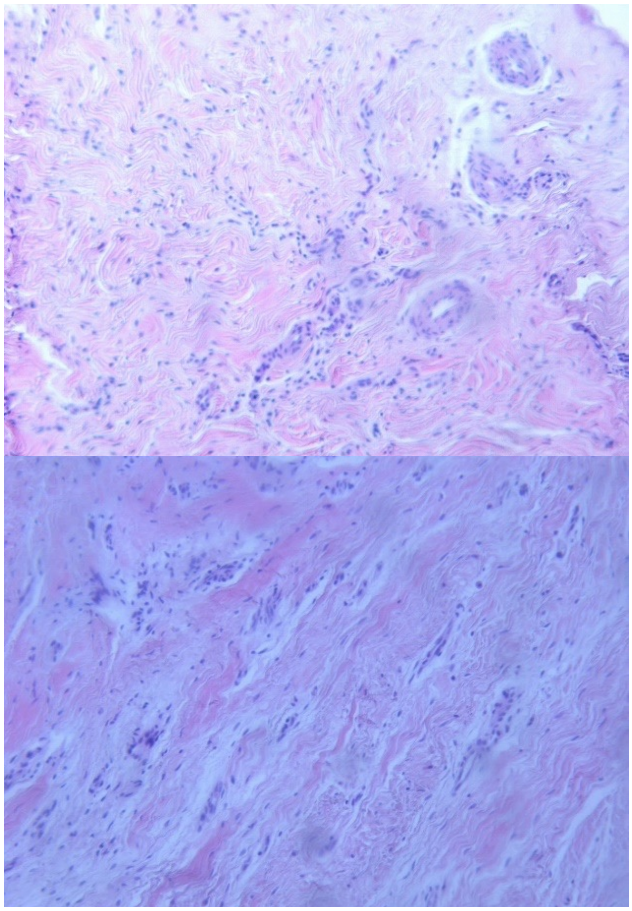

Figure 5. Histologic image of inflammatory myofibroblastic tumour showing proliferation of fibroblasts and myofibroblasts, accompanied by mixed inflammatory infiltration by plasma cells, lymphocytes, eosinophils and histiocytes.

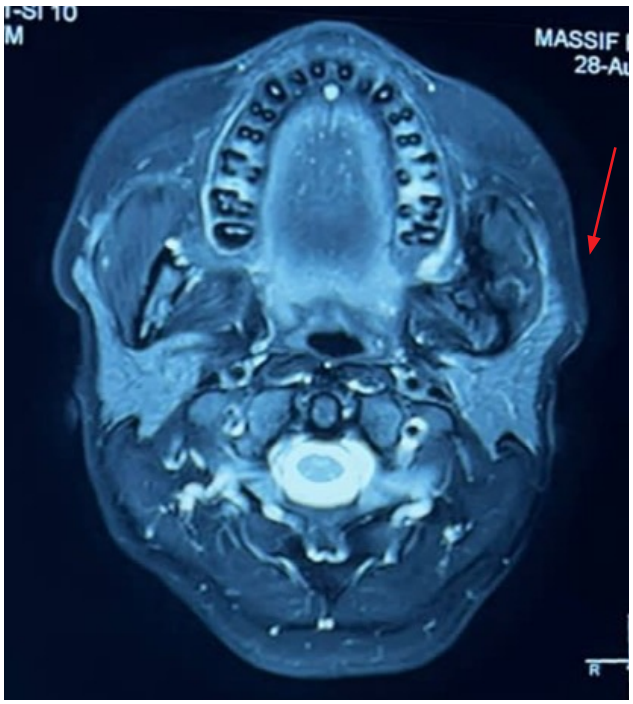

Figure 6. The stable aspect of IMT in the ascending branch of the mandible.

On contrast-enhanced CT, IMTs are distinguished as homogeneous or heterogeneous lesions, with variable enhancement due to the presence of fibrosis. On gadolinium contrast-enhanced MRI the lesion appears with variable enhancement. On T1-and T2 weighted sequences, IMTs usually show low signal intensity, also reflecting the presence of fibrotic tissue[6]. CT and MRI are the reference techniques to evaluate these tumours.

IMT is classified by the World Health Organisation as having intermediate biological potential, mostly due to a tendency for local recurrence. Multiple sources have reported a recurrence rate of 25% for extrapulmonary lesions. The mainstay of treatment is surgical resection; however, it can prove difficult depending on the location and extension of the lesion, and the risk of mutilation. This is the case in our patient. For unresectable lesions, the use of targeted molecular therapy has been utilised and proved to be efficacious.

Kube et al. collected a cohort of 38 patients with IMTs with heterogeneous courses of variable treatments and showed that chemotherapy with alkylator-based regimens seems to be justified to attempt to reduce tumour size and enable tumour resection[7].

Corticosteroid therapy reduces the inflammatory component of the tumour. In this case, we obtained an improvement of clinical signs, but the tumour remained stable. Combined use of ALK and COX-2 inhibitors as adjuvant therapy are promising in the treatment of head and neck IMTs. Radiotherapy is rarely discussed and its effects seem controversial in the literature[7].

Regarding the prognostic factor, Zhang et al. conducted a retrospective study of 41 patients presenting head and neck IMT and demonstrated that 66.7% of lesions had a moderate or severe degree of inflammation more than those in other sites. This is due to the location being adjacent to the upper aerodigestive tract, and the abundance of lymphatic tissue[8]. Among an abundance of inflammatory factors, COX-2 may play a role and its mechanism is promoting cell proliferation, inhibition of cell apoptosis and accelerating angiogenesis.

The best prognoses have been documented following radical surgical resection with negative margins, which is curative in over 90% of extrapulmonary localisations, including the head and neck[5]. They have a low rate of local recurrence and dissemination[9]. The response of IMTs to corticosteroids has been well documented, however, worsening of IMTs as well as reoccurrences following steroid treatment have been observed[10]. In our case, the patient remained corticosteroid dependent, and is taking this treatment regularly for a better prognosis.

CONCLUSION

IMT is a challenging disease. Given its variable clinical and radiological presentation, it is of paramount importance to be aware of it so it can be swiftly recognised for a diagnosis to be promptly made. The adapted treatment should be started immediately to prevent possible life-threatening outcomes.

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