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Inflammatory myofibroblastic tumor in the maxillary sinus with asymptomatic course excised during endoscopic surgery – a case report and review of the literature

Zapalny guz miofibroblastyczny zatoki szczękowej o bezobjawowym przebiegu usunięty podczas operacji endoskopowej – opis przypadku i przegląd literatury

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ABSTRACT:

Introduction: Inflammatory myofibroblastic tumor (IMT) is a rare condition that can mimic potentially more dangerous states such as malignant tumors. The tumor itself can also show local malignancy as well as malignant transformation. The paranasal sinus IMT is quite a rare case in the literature. The manifestation of the disease can include face swelling, nasal obstruction, epistaxis, vision acuity worsening, numbness of the face, pain. Etiology of this type of lesion still remains uncertain but there are a few assumptions on the issue: viral and genetic, as well as posttraumatic and postinflammatory. We report a case of an adult woman with IMT detected in the right maxillary sinus after endoscopic sinus surgery.

Case report: We report the case of an adult woman with IMT detected in right maxillary sinus after endoscopic sinus surgery.

KEYWORDS: endoscopic sinus surgery, inflammatory myofibroblastic tumor, inflammatory pseudotumor of the maxillary sinus, plasma cell granuloma

STRESZCZENIE:

Wstęp: Zapalny guz miofibroblastyczny (IMT) jest rzadkim stanem, który może naśladować potencjalnie bardziej niebezpieczne stany, takie jak nowotwory złośliwe. Sam nowotwór może również wykazywać miejscową złośliwość, a także złośliwą transformację. IMT zatoki przynosowej nie występuje często w literaturze. Objawem tej choroby mogą być: obrzęk twarzy, niedrożność nosa, krwawienie z nosa, pogorszenie ostrości wzroku, drętwienie twarzy, ból. Etiologia tego typu zmian nadal pozostaje niepewna, ale istnieje kilka założeń w tym zakresie: wirusowych i genetycznych, a także pourazowych i pozapalnych.

Opis przypadku: Niniejszy opis przypadku dotyczy dorosłej kobiety z IMT wykrytym w prawej zatoce szczękowej po endoskopowej operacji zatoki.

SŁOWA KLUCZOWE: endoskopowa operacja zatok, zapalny guz miofibroblastyczny, zapalny guz rzekomy zatoki szczękowej, ziarniniak plazmatycznokomórkowy

ABBREVIATIONS

ALK – anaplastic lymphoma kinase
CT – computed tomography
EBV – Epstein-Barr virus
HIV – human immunodeficiency virus
IMT – inflammatory myofibroblastic tumor
MRI – magnetic resonance imaging
ROS-1 – receptor tyrosine kinase

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) (also known as inflammatory pseudotumor, fibrous xanthoma, plasma cell granuloma, pseudosarcoma, lymphoid hamartoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, benign myofibroblastoma) [1] is a rare condition that can mimic potentially more dangerous states such as malignant tumors. The tumor itself can also show local malignancy as well as malignant transformation,

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but available literature reports such situations as "hardly possible". A resected lesion constitutes a circumscribed mass with a rubbery to firm cut surface [2]. Microscopically, it consists of smooth muscle cells, connective tissue components and immune cells in various proportions. Its common locations include lungs, mesentery, bladder, but also heart, temporal bone and intracranial space. The paranasal sinus is an extremely rare site [3, 4]. We report a case of an adult Caucasian woman with an inflammatory myofibroblastic tumor detected postoperatively in the right maxillary sinus.

CASE PRESENTATION

A 54-year-old Caucasian woman was admitted to the department of otorhinolaryngology due to a mass found in CT (computed tomography) of her right maxillary sinus. The lesion was revealed incidentally about 3 months earlier, during the diagnostic process of a painful lesion on the forehead, which drained spontaneously. The lesion did not cause any bone destruction.

At the visit she presented with chronic rhinosinusitis symptoms, i.e. mucosal swelling, mucosal hypertrophy of the right and left inferior turbinates. However, she did not report any pain of her face, nasal obstruction, blockage, discharge or purulence, postnasal drip or hyposmia. There was no history of nicotinism. However, she reported recurrent bronchitis.

The CT showed that the right maxillary sinus was filled completely with a soft-tissue lesion and dense fluid. Besides, a periosteal reaction was present. Minor polypoidal mucosal thickening in the right part of the sphenoid sinus was detected. Moreover, there was a concha bullosa variant of the right middle turbinate and nasal septum deviation to the left. Other paranasal sinuses were intact. There was no bone infiltration by any process. Taking into consideration the CT results as well clinical presentation, the differential diagnosis of chronic paranasal sinusitis or inverted papilloma was suggested.

The results of the physical examination, as well as no middle nasal meatus involvement argued for the first entity. The latter was considered because of asymmetrical impact of the lesion on sinuses, especially the maxillary sinus. Having evaluated the CT results, a decision on complete mass resection during endoscopic sinus surgery was taken. Biopsy was not considered as a diagnostic option, as the whole lesion was in the sinus, not protruding from the semilunar hiatus.

The operation was performed under general anesthesia, with the 30- and 70-degree view angle rigid sinuscopes. The surgery covered the following: right-sided uncinectomy, middle turbinate partial remodeling and a wide antrostomy. The solid mass filling the whole maxillary sinus space was completely extracted macroscopically with rigorous sinus mucosa debridement. Its diameter was ca 2.5 cm. The inferior turbinate was also removed. No complications occurred in the perioperative period. The microscopic analysis of the biopsy revealed IMT in the right maxillary sinus and rhinitis chronica in the inferior turbinate.

The postoperative follow-up visits took place once a week during the first month, then once a month. One month after surgery, nasal

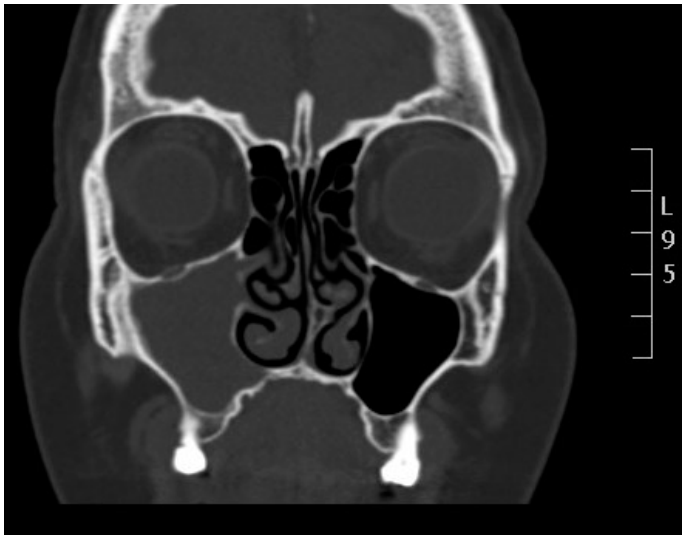

Fig. 1. The computed tomography scan of the paranasal sinuses, coronal reconstruction. Soft tissue mass in the right maxillary sinus, without bone destruction patterns.

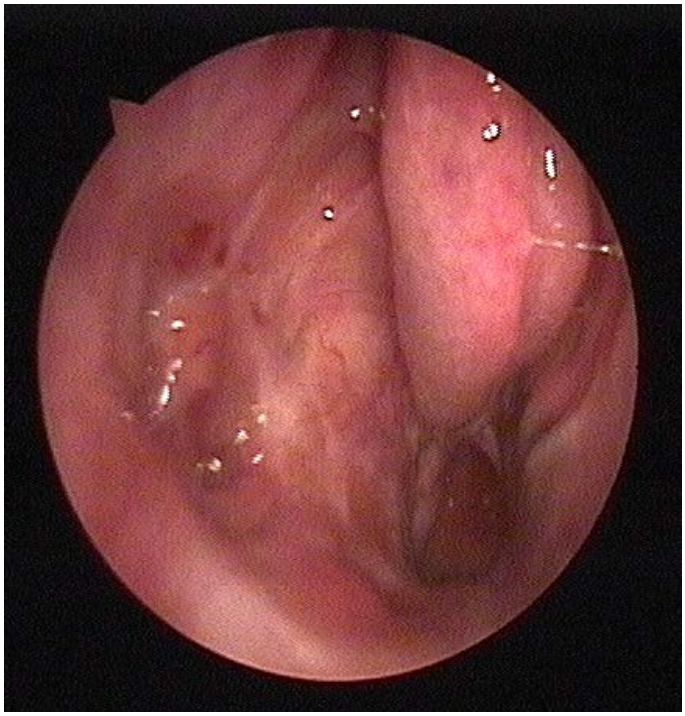

Fig. 2. The right maxillary sinus 4 months after surgery with no recurrence visible – an endoscopic view.

corticosteroids were prescribed as rhinosinusitis symptoms were detected during regular follow-up examinations. No pattern of tumor recurrence was observed for over 4 months after surgery.

DISCUSSION

Inflammatory myofibroblastic tumor is defined by the World Health Organization as ‚a distinctive lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils. It occurs primarily in soft tissue and viscera of children and young adults' [5]. This pathology was first

described in 1939 by Brunn in lung's specimen (on the other hand, some authors claim that 'idiopathic orbital inflammatory syndrome' described by Birch-Hirschfeld in 1905 [6] refers to IMT) [7]. Since then, many other locations of the tumor have been reported on, but still the paranasal sinus IMT is quite a rare case in the literature. The reason could be misdiagnosis as low incidence of this tumor makes pathologists being unfamiliar with its microscopic characteristics. The second cause might be asymptomatic course in a vast majority of this type of lesions, as in the case of the reported patient. Due to this fact, patients tend not to visit otorhinolaryngologists. However, the above-mentioned hypotheses seem to be difficult to proof due to a low number of described cases.

The symptomatology depends on the site involved. In our case there were no certain signs or symptoms which could have suggested the progression of the tumor in the maxillary sinus, albeit literature gives many examples of a potential clinical presentation when paranasal sinuses are the primary site of the mentioned illness. This includes face swelling, nasal obstruction, epistaxis, vision acuity worsening, numbness of the face, pain (also referred). (Nota bene, other locations of IMT can have such systemic symptoms as fever or weight loss).

The etiology of IMT still remains uncertain but there are a few assumptions on the issue: viral and genetic among others, as well as posttraumatic and post inflammatory.

Human herpesvirus 8 DNA sequences have been found in some cases [8]. Other possible infectious agents include *Helicobacter pylori*, *Pseudomonas*, *Escherichia coli*, *Klebsiella*, *Bacillus*, *Mycobacterium*, *Actinomycetes*, *Nocardia*, *Mycoplasma*, HIV, EBV [1]. Subsequently, some serology techniques and/or genetic material identification methods may be of help.

The genetic finding in most IMT cases is overexpression of the anaplastic lymphoma kinase (ALK) gene. The product of this gene is responsible for embryonic nervous system development but was first described as a protein expressed in the majority of anaplastic large-cell lymphomas [9]. However, also diffuse large B cell lymphomas and rhabdomyosarcomas present ALK-positive staining. Some researchers suggest a predictive role of ALK staining in IMT. Chun et al. small population study on children showed that ALK expression and complete surgical resection were markers of good prognosis [10]. ALK-negative lesions could be more prone to form distant metastasis [11].

Computed tomography and magnetic resonance imaging seem to be the first-line option of visualization of IMT, though the second one is considered to be a better visualization technique to reveal nerve and muscle involvement [12]. It usually presents as a solid mass, with a nonuniformly enhanced signal. Infiltration of vicinal structures can take place in a more severe course. Low intensity on T2-weighted images is characteristic for fibrotic composition [13].

Cheng K.J. et al. study, based on cases of 8 patients with neck IMT, names the following patterns of this entity: I) A soft-tissue density, rarely exhibiting calcification or necrosis on CT scans; II) when enhanced, the mass displays enhancement on CT and MR images; III) MRI is superior to CT scans in the differential diagnosis of this disease; IV) as this tumor often has multiple components, it usually presents with heterogeneous signals; V) in general, the lesion displays a hypointense-isointense signal on T1-weighted sequences and an isointense-hyperintense signal on T2-weighted sequences; VI) due to the fibrous tissue in the tumor, delayed enhancement may be observed on gadolinium-enhanced MR images; and VII) due to its benign or intermediate features, the tumor is usually a well-defined mass [14]. Angiography is not considered the basic imaging method of these tumors. It was rather performed in cases where a juvenile angiofibroma had been suspected [15, 16]. Nonetheless, it can reveal the hypervascularity of the tumor, blood supply by mildly hypertrophied terminal branches of the internal maxillary artery. Attenuated tumor staining with slow washout during the capillary phase might be observed [15].

The histological patterns of IMT are frequently categorized into 3 or 4 groups. According to the Coffin's classification [17], these are: 1) hypocellular pattern with inflammatory cells dominant, with the myofibroblasts arranged loosely; 2) hypercellular – a compact fascicular proliferation on a collagenized background; 3) loose spindle-cell arrangement resembling scar tissue. Among various authors there is a consensus that the treatment of choice should be a radical surgical excision whenever possible. In our patient we carried out fully endoscopic surgery to excise this lesion, mainly due to other possible diagnoses. Other options include radio- or/and chemotherapy. Hansen C. et al. reported diminishing tumor mass after intensity-modulated radiotherapy followed by ifosfamide, dacarbazine, and celecoxib treatment [18]. Anti-inflammatory drugs, steroid as well as non-steroid, were used with good effect [19, 20].

Patients with revealed ALK rearrangement can respond to treatment with crizotinib, small-molecule inhibitor of the receptor tyrosine kinases ALK and ROS-1, which is registered to treat metastatic non-small cell lung cancer with ALK- or ROS-1-positive tests [21]. To the best of our knowledge, there still does not exist any predictive pattern, showing which drugs would be the most efficient in particular IMT patients. As mentioned above, polypragmasia was used in an attempt to reduce tumor mass. The prognosis for IMT is generally benign, as no important organs are involved with the disease. Even so, a detailed examination should be performed at any follow-up visit due to possible recurrence and metastases (about 25% and <5% respectively in an extrapulmonary location) [17].

CONCLUSIONS

The inflammatory myofibroblastic tumor is quite a challenging type of neoplasm for clinicians, surgeons and pathologists. The tumor is still poorly recognized. Due to its rare occurrence, especially in paranasal sinus location, our is an attempt of extrapolation of the knowledge about IMT in this particular region, with all advantages and disadvantages. Our case report shows that IMT can be a very diagnostically difficult entity, because of its asymptomatic development. Endoscopic surgery was performed in the abovementioned patient with no tumor recurrence in a short follow-up period. Further studies are needed to evaluate if this can be the first-line treatment in the maxillary sinus IMT.

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