Inflammatory Myofibroblastic Tumor Arising in the External Ear: Unexpected Location. (Case Report)

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Abstract:

We present an unexpected extremely rare case of an inflammatory myofibroblastic tumor (IMT) of the external ear. A 3-year-old boy presented with a mass arising on the posterior aspect of the left ear lobule and extending to the external auditory canal. This mass grew slowly and showed no skin changes. Radiologically, it was diagnosed as a vascular tumor. Surgical excision and primary closure was performed for the patient. Histopathologic examination demonstrated a circumscribed noncapsulated mass formed by a proliferation of spindle cells in deep dermis and subcutaneous tissue. The spindle cells were mixedwith lymphoplasmacytic infiltration compatible with the IMT. Because it is very rare to arise in this location, we present this case as a case report with details histopathological examinationand reviewed the English literature for similar cases.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a disease were first observed in the lung and described by Brunn in 1939. [1,2] Then, it was named as inflammatory pseudotumor by Umiker et al in 1954 because it mimics malignant neoplasms clinically, radiologically, and microscopically. [1] It has many other names also as plasma cell granuloma, plasma cell pseudotumour, inflammatory myofibrohistiocytic proliferation, omental mesenteric myxoidhamartoma, and inflammatory pseudotumour. [3-6] The nature of this disease is still controversial, some pathologists consider it as an inflammatory reactive process (based on its plasma cells polyclonality, mixed population of cells, and absence of atypia) whereas others consider it as a neoplastic one (based on its association with ALK gen re-arrangement on chromosome 2p23, its recurrence, and metastasis). [5, 7] This controversy makes the definition of IMT a little bet difficult. According to the World Health Organization (WHO) 2002, IMT is defined as "*a distinctive lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils*". [3] It is an uncommon disease. There is no single defined cause for it but it is known that IMT is associated with trauma and immunosuppression status. [2-3]The later have been observed recently by its association with human herpes virus-8 (HHV-8). [3] It formed by a spindle fibroblastic and myofibroblastic cells that arranged in fascicles and storiform patterns with myxoid background reach in vessels and mix inflammatory cells. It may show some mitosis but not atypical one.It is usually arising in children and young adults, but no age group is immune from it. [3] The clinical presentation is variable and depends on the site of involvement. [2, 5] By far, the commonest location for this lesion is the lung and the patient will develop shortness of breath in association with some constitutional signs and symptoms as fever, weight loss, and elevated ESR. [3] Among the extrapulmonary IMT, 43% of the cases are arising in the mesentery and omentum. Other sites include soft tissue, mediastinum, GIT, GU tract, and head & neck regions. Head and neck IMT accounts only 5% of all cases of IMT and the orbit site been the most frequently involved site. [2, 5-6, 8-9] It is extremely rare to arise in the ear as a primary site of origin. For this reason and due to no enough review of literature about this disease in this particular location, we report this case as a case report with details histopathological examination and reviewed the English literature for similar cases.

Case Report

The patient was a 3-year-old boy who had presented with left ear mass. This mass was arising on the posterior aspect of the ear lobule and extending to the external auditory canal. It was lobulated and covered by a congested skin without any ulceration. There was no hearing loss or any neuronal deficiency. Rest of ear and systemic examination was unremarkable. Computed tomography (CT) and magnetic resonance imaging (MRI) showed irregular and lobulated mass measured 3 x 1.5 x 1.5 cm. It showed intense enhancement after intravenous contrast administration. There was a small extension into the adjacent mastoid with focal erosion of the cortical bone. No intracranial extension was noted. Theother parts of the ear were unremarkable. According to these radiological features, the differential diagnosis included: hemangioma, hemangiopericytoma, and glomus tumor. The tumor have been surgically resected and sent to the histopathology lab. On grossing, the specimen was received in 10% neutral bufferedformalin and consisted of multiple pieces of tan tissue covered partially by unremarkable skin. They were measuring in aggregates 1.2 cm in maximum dimension. The specimen is submitted entirely in one cassette for histopathological examination. After 16 to 18 hours of fixation, the specimen has been processed according to the standard procedure (first, dehydration by different concentrations of alcohol, then clearing of alcohol by xylene, and lastly infiltration of the tissue by paraffin). After that, the tissue is embedded in a labeled paraffin block. By using the microtome, 4 micron thickness section from this block is made. This section was shifted to a glass slide in order to stain it with hematoxylin and eosin stains (H&E) according to the standard protocol.Microscopic examinationof this slide demonstrated a circumscribed noncapsulated dermal and subcutaneous lesion formed by spindle cells. These cells were mainly fibroblasts and

myofibroblasts with a highly vascularized myxoid background that contains many mixed inflammatory cells (figures 1 and 2). There were some mitotic activities (4/10 hpf). Focal mild atypia was noted. No calcification was seen. Immunohistochemistry (IHC) staining showed that these cells were positive for smooth muscle actin (SMA) (figure 3), caldesmon (figure 4), and ananplastic lymphoma kinase (ALK) (figure 5). They were negative for desmin, muscle specific actin (MSA), CD34, and S100.According to the histopathological features as well as the immunohistochemicalresults, a diagnosis ofinflammatorymyofibroblastic tumor(IMT) was made.

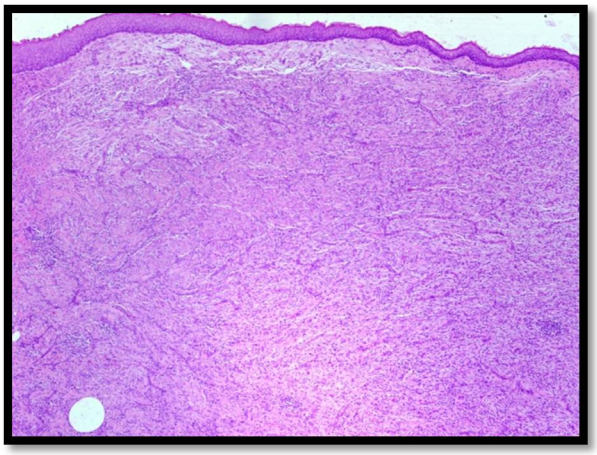

Figure-1: Low power demonstrates that the lesion is formed by spindle cells in a background of mixed inflammatory cells (H&E x40)

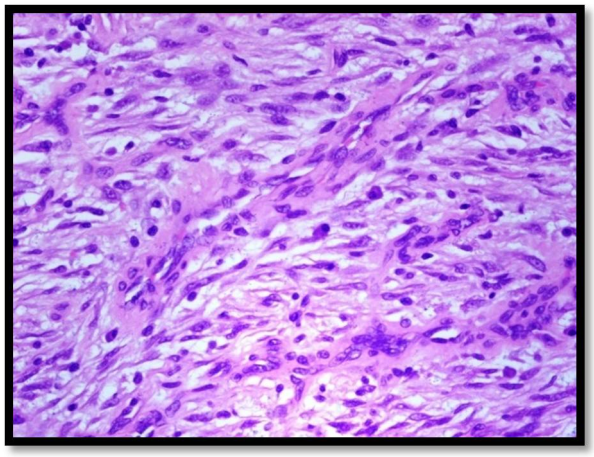

Figure-2: High power shows spindle fibroblasts and myofibroblasts with a myxoid background rich in mixed inflammatory cells, mainly lymphocytes and plasma cells (H&E x400).

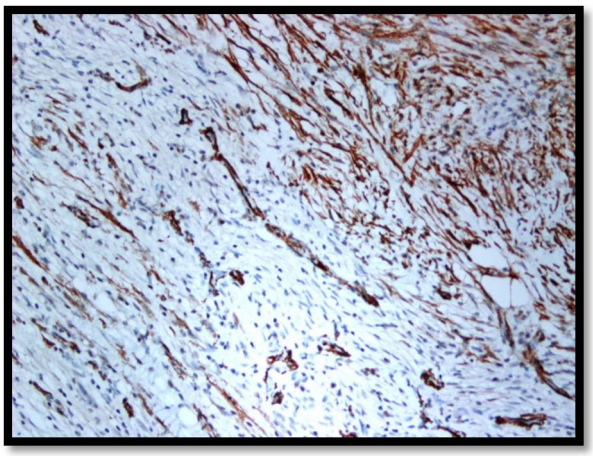

Figure-3: The spindle cells are focally positive for smooth muscle actin (SMA) (immunohistochemistry stain x200).

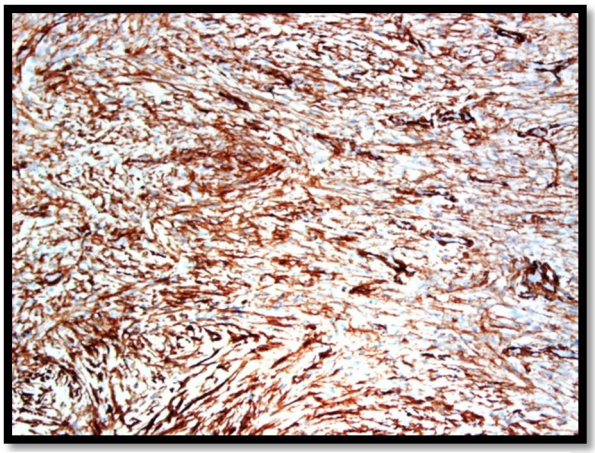

Figure-4: The spindle cells are diffusely positive for caldesmon. (immunohistochemistry stain x200).

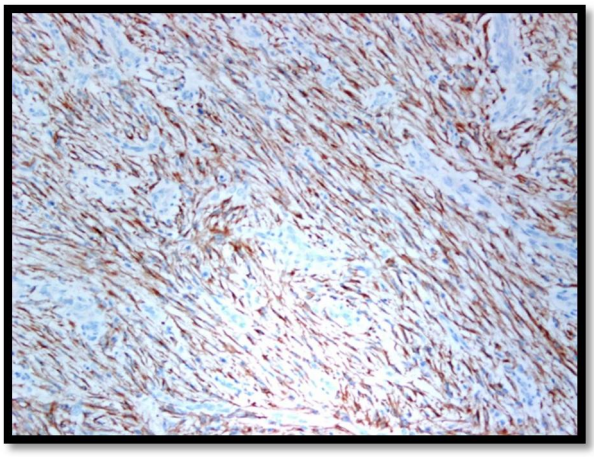

Figure-5: The spindle cells are diffusely positive for anaplastic lymphoma kinase (ALK). (immunohistochemistry stain x200).

Discussion

Generally speaking, the vast majority of IMT cases are arising in the pulmonary area [3] and most of extrapulmonary cases occur in the mesentery and omentum. According to H.S. Ong et al study, ear IMT cases represents only around 7% of head & neck caseswhich represent 5% of all IMT cases. [10] By reviewing the English literature using the National Library of Medicine PubMed online database (http://www.ncbi.nlm.nih.gov/pubmed/) with the keywords "*inflammatory pseudotumor ear*" and "*inflammatory myofibroblastic tumor ear*", we found only 8 cases of IMT involving the ear as the main primary site of origin(and our case become the 9th). The age of the patient in those cases is ranging from 2.5 year to 55 years old. Based on the gender of the patients, the cases can be divided into 5 males and 4 females. For unknown reason, the vast majority of these cases (including our case) are arising from the left ear. Most of auditory IMT cases are arising in the middle ear (4 cases). Only a single case is arising from the inner ear. The lesion of our case was arising at the ear lobule and extending to the external auditory canal. The main presenting symptoms of the cases that arising from the external ear is painless mass without any hearing or neuronal problems, as in our case. The speed of growth is vary, some grow rapidly mimicking malignancy, [2] others grow slowly like our case. [9] The reason for this variability in the growth speed is not clear. Cases that arising from the middle and inner earshave different kind of symptoms including hearing loss, otalgia, and facial nerve paralysis. [4, 6-8, 11]The sizerange of these cases is from 1cm to 4cm in maximum dimension. Our case is still within this range (3cm based on the radiological studies).

On radiological examination, those cases show soft tissue masses with irregular ill-define border. Some of those cases show infiltration and destruction to the adjacent boney structures. It may show intravenous contrast enhancement, as in our case. [4, 8] Generally speaking, there is no specific radiological sign for this lesion. [2] The differential diagnosis based on the radiology alone includes cholesteatoma, skull base osteomyelitis, neoplasms (paragangliomas, facial neuromas, hemangiomas), and granulomatous disorders (e.g., Wegener's granulomatosis and Langerhans cell histiocytosis). [2, 4, 8] In our case, vascular tumors were the main differential diagnosis, which can be explained by the high vascularity of the lesion under the microscopic examination.

The accurate diagnosis for such a lesion is made mainly by the pathologists in the histopathology lab. The gold standard type of biopsy in those cases is incisional/excisional biopsy. [10] Core biopsy is the minimal requirement to make this diagnosis. [10] Fine needle aspiration (FNA) sample is inadequate to reach the diagnosis because mostly it will provide a hypocellular material that is inadequate for examination. [9-10] Intra-operative frozen section consultation should be avoided as much as possible because it will be a misleading to the surgeon during the operation. [8, 11] The pathologist should not make a specific diagnosis for any spindle cell lesion unless he/she is 100% sure from the diagnosis because different types of spindle cell lesions have different approach for treatment [8, 11] that may have an unrepairable consequence. For example, the treatment of IMT should be complete excision with free surgical margin but without removing any nerve whereas in facial neuroma, the treatment will include removing the nerve and put a graft instead of it. This may lead to some unnecessarily neuronal abnormalities.

On the microscopic examination, almost all the cases share the same morphological picture. The tumor shows a relatively circumscribed noncapsulated dermal and subcutaneous lesion formed by spindle cells arranged in fascicular and/or storiform patterns. These cells are mainly fibroblasts and myofibroblasts with a highly vascularized myxoid background. The background also contains many mixed inflammatory cells, mainly lymphocytes and plasma cells. There are some mitotic activities but no atypical one is seen. No/mild focal nuclear atypia can be present. Immunohistochemical (IHC) staining shows that these spindle cells are positive for myofibroblastic lineage as smooth muscle actin (SMA), and caldesmon. Some but not all the cases are positive for anaplastic lymphoma kinase (ALK-1), which is a helpful stain to make such a diagnosis but it is associated with worse prognosis. [8, 12] They are negative for desmin, CD34, and S100.

The treatment of auditory IMT is still controversial due to the rarity of these kinds of cases in this location [3, 10-11] but according to H.S. Ong et al, surgical resection with negative surgical margins is the gold standard treatment for head and neck IMT cases because it associated with the greatest likelihood of cure. [8] If the lesion is not completely removed, there is 50% chance to the lesion to locally recur. [8, 10] Others physician suggest the use of systemic cortisone or non-steroidal anti-inflammatory drugs as a medical treatment for those incompletely excised cases. [8] Radiotherapy and chemotherapy have been used in aggressive IMT cases. [8]

Conclusion

In conclusion, we present an extremely rare location of IMT involving the external ear of a child. The presenting symptoms will be determined by the specific site of involvement in the ear. Preoperative diagnosis of this IMT was difficult due to its rarity and the lack of specific findings. Incisional/excisional biopsy is the gold standard specimen to make such a diagnosis. FNA and intra-operative frozen section consultation should be avoided in such cases. The treatment of choice of such cases is still controversial.

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