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Computed Tomography of Infantile Myofibroma in the Ethmoid Sinus: A Case Report

We report a 4-year-old girl complaining of diplopia and a small lump in the medial side of the left orbit. CT scan showed a mass at the anterior ethmoid sinus with erosion and expansion. The mass was excised and the diagnosis was solitary infantile myofibroma (IM).

Keywords: Solitary Infantile Myofibroma, Ethmoid Sinus, Computed Tomography

Introduction

Myofibroma and myofibromatosis are benign neoplasms consisting of myofibroblastic cells, often in the dermis or submucosal structures. Myofibroma, which is the single form, is most common in the head and neck. As its name suggests, infantile myofibroma (IM) is a pediatric disorder with over half of the cases showing up at birth and nearly 90% during the first two years of life.[1] Although IM is the most common fibrous tumor of infancy, many consider it as a rare disease. Its presentation in an older child or adult is even more outstanding.[1,2] We present an unusual case of IM in an older child. Among 307 cases of oral spindle cell neoplasm, myofibroma was seen in eight cases.[3]

In the literature, several classifications have been described for pediatric fibromatosis of which Enzinger and Weiss's categorization is the most widely accepted. The desmoid fibromatosis are locally aggressive, recurring lesions, typically occuring in the extremities or the trunk and similar to the fibromatosis found in adults. In contrast, myofibromas are in general, characteristic of infancy and childhood and they have little or no clinical or morphological equivalents in the adult life.[4,5]

Myofibromatosis was first described in 1954, by Stout[6] as "congenital generalized fibromatosis", a term he chose to define a disseminated disease of multiple nodular lesions in the newborn. Stout also recognized fibromatosis from fibrosarcoma, although locally invasive, multicentric, and often aggressive. For many years, the standard treatment of fibromatosis of all types was aggressive, wide, local resection, based on a review of 40 cases of fibromatosis, including 10 children younger than 15 years by Conley et al.[4] Later, Chung and Enzinger argued for the classification of infantile myofibromatosis as a discrete lesion, based on its unique clinical and tissue staining characteristics consistent with myofibroblastic origin.

Case Presentation

We report a 4-year-old girl with a 6-month complaint of diplopia and a small

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lump in the medial side of the left orbit. CT scan was performed (Figs. 1A&B).

In the CT scan imaging, there was a round mass lesion with soft tissue density (43-hunsfield unit) without calcification at the left side of the anterior ethmoidal sinus with erosion and expansion of lamina papyracea and lateral deviation of the left globe. It started from the cribriform plate and ended at the level of the middle turbinate.

Total excision of the mass was carried out. It was a 30×25×35mm mass with an external nodular surface. Microscopically, it showed ovoid cell proliferation with cigar shaped nuclei devoid of cytological atypia arranged in wide bundles associated with spindle cells and scattered lymphoplasmacytic infiltration (Fig. 1C&D). The immunohistochemical profile revealed vimentin positive, desmin weakly positive, S100 focally positive and smooth muscle actin (SMA) strongly positive. The diagnosis was solitary infantile myofibroma.

Follow-up showed no recurrence after 3 months.

Discussion

IM is a rare mesenchymal disorder characterized by benign myofibroblastic tumors and is considered as the most common fibrous tumor of infancy.[7] IM appears in three different types: solitary lesions, multi-

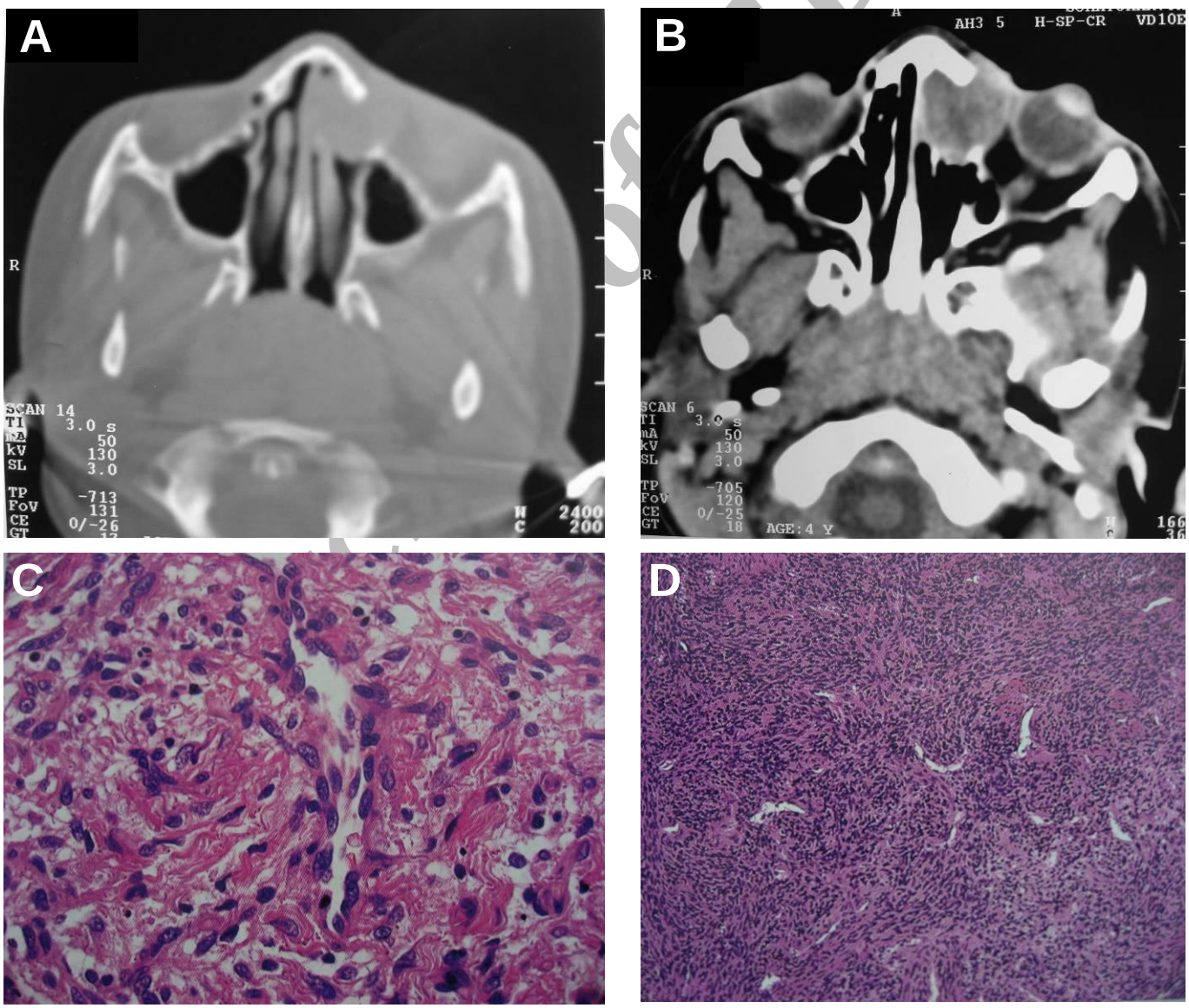

Fig. 1. A 4-year-old girl with infantile myofibroma.

A & B. Axial CT scan shows a mass in the medial side of the left orbit with erosion and expansion to the ethmoid sinuses.

C & D. Ovoid cell proliferation with cigar shaped nuclei associated with spindle cells.

centric lesions without involvement of viscera and multicentric lesions with involvement of viscera. As its name suggests, it is considered a pediatric disorder with approximately 60% of cases presenting at birth or during the perinatal period and 80-88% by 2 years of age.[1] However, there are isolated reports in the literature of IM occurring in older children and adults.[7]

Based on the review of 130 patients with a diagnosis of head and neck neoplasm in Beck et al.'s study[4] from 1975 to 1997, 13 patients had fibromatosis and only one of them had ethmoidal myofibromatosis.

The list of differential diagnoses for IM includes fibrosarcoma, fibrous histocytoma, desmoid tumors, leiomyosarcoma and in the newborn, delivery trauma to the sternocleidomastoid muscle. Although clinical evaluation includes imaging studies, as IM does not have a characteristic appearance on CT or MRI, a definitive diagnosis is not possible. In addition, the radiographic appearance of IM, particularly on MRI, may be misleading, as a result of aggressive appearing and infiltrating lesions. However, imaging can be helpful in evaluating the progression, regression, recurrence and the extent of the disease. Plain films for bony lesions demonstrate well-circumscribed osteolytic masses with sclerotic margins. CT images show heterogeneous appearance, cystic or homogeneous areas, bony remodeling and calcification.[4,7]

The lesions are hyperintense, isointense or hypointense to brain parenchyma in T1 and T2 weighted images of MRI. However, contrast enhancement is seen in IM lesions in both CT and MRI imaging.[7]

In our patient, a mass was detected in the ethmoidal sinus with an almost homogeneous density and bony erosion without calcification.

The microscopic appearance of fibromatosis tumors does not often reflect the generally benign clinical characteristics. Histological findings such as local invasion, which indicate malignancy, are not uncommon. Metastasis and malignant degeneration have not been reported. Enzinger[5] mentioned the higher aggressiveness and higher therapy resistance of highly cellular lesions in infants compared to the less cellular or fibrotic lesions, which were more differentiated and clinically, more benign. Other reports suggest that the interpretation of the biopsy specimens varies widely on the portion of the tumor sampled. Indeed, the pathological distinction between fibromatosis, aggressive fibromatosis and fibrosarcoma may be challenging, even for experienced pathologists.

The morbidity of fibromatosis depends on the location of the lesion. Some lesions may not be diagnosed if they relapse without causing symptoms.

Conservative surgical eradication is advised for tumors affecting vital functions or causing growth abnormalities.[5]

References

1. Coimbra F, Moreira P, Lopes O. Solitary myofibroma in the oral vestibule. Rev Port Estomatol Cir Maxilofac 2008 Feb;49(2):93-5.
2. Chung EB, Enzinger FM. Infantile myofibromatosis. Cancer 1981 Oct 15;48(8):1807-18.
3. Jordan RC, Regezi JA. Oral spindle cell neoplasms: a review of 307cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2003 Jun;95(6):717-24.
4. Beck JC, Devaney KO, Weatherly RA, Koopmann CF Jr, Lesperance MM. Pediatric Myofibromatosis of the head and neck. Arch Otolaryngol Head Neck Surg 1999 Jan;125(1):39-44.
5. Enzinger FM, Weiss SW. Fibrous tumors of infancy and childhood. In: Soft tissue tumors. 3rd ed. St Louis, MO: Mosby-year book; 1995. p. 231-68.
6. Stout AP. Juvenile fibromatoses. Cancer 1954 Sep;7(5):953-78.
7. Franzese CB, Carron J. Infantile myofibromatosis: unusual diagnosis in an older child. Int J Pediatr Otorhinolaryngol 2005 Jun;69(6):865-8.