A case of Juvenile psammomatoid ossifying fibroma in the ethmoid sinus

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Discussion
Juvenile psammomatoid ossifying fibroma (JPOF) is a very rare disease that is classified into ossifying fibroma. Histologically it is benign but shows local invasiveness. It tends to occur in facial bone. CT scan shows high density punctate shadow which indicates osteoid. However, it is difficult to distinguish it from fibrous dysplasia by this viewpoint. Pathological diagnosis is essential. Typically MRI shows from intermediate to low signal on T1-weighted scan and intermediate to high signal on T2-weighted scan. Intracranial invasion, narrowing of jugular vein and the carotid artery are important points. The first-line therapy is complete remove by operation because it shows 30-50% chance of reoccurrence. As far as we searched, there is no report of malignant transformation and we can also expect vanishing of tumor with aging. Careful Regular follow up is required.

Case: 7 year-old boy

History of present illness
Left ophthalkocele continued 2 months and he first visited the department of ophthalmology. CT and MRI of orbits revealed a tumor in the left ethmoid sinus, therefore, the patient was referred to our department.

Findings and images
Exophthalmos (+) double vision (-) visual field defect (-) biochemical examination of blood: within normal limit
The tumor was 38 × 24mm in size. The tumor compressed surrounding bone, and displaced only the left eye-orbit lateral wall. (figure1,2)

Treatment progress
Endoscopic tumorectomy under general anesthesia was performed for the purpose of diagnosis and treatment. The tumor was elastic and hard, surrounded by an ossifying septal structure. Inside of the tumor was hemorrhagic. We only removed ossifying septum of the tumor and left the septum of the tumor at the orbit side to avoid the excessive bleeding. After the operation ophthalmologic symptom disappeared immediately.

Treatment Plan
regular follow up by MRI except the case of functional disorder, cosmetic problem, ocular and intracranial complication

Postoperative course
6 months after the first operation
MRI showed solid tumor have changed to liquid component
There was no tendency of tumor enlargement.
12 months after the first operation (fig3)
MRI indicated that tumor showed enlargement in size → palliative reductive surgery
42 months after the first operation (fig4)
MRI showed enlargement again → palliative reductive surgery
He underwent 3 times operation in total and the tumor still remains.