

Case Report

Enchondroma of the Skull Base in a Case of Ollier's Syndrome

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ABSTRACT

Ollier's syndrome, a variant of multiple enchondromatosis, is a rare disease with an estimated prevalence of 1/100,000, characterized by multiple enchondromas, asymmetrically involving small bones of the hands and feet, especially the proximal phalanges. Intracranial enchondromas, such as those arising from the skull base are extremely rare. Herein, we report a 25-year-old female, known case of Ollier's disease, presenting with right eyelid ptosis and visual disturbance. Brain MRI revealed a skull base tumour suspicious to enchondroma followed by trans-sphenoidal resection. Histologic examination of the excisional biopsy sample confirmed the diagnosis of enchondroma.

Keywords: Enchondroma, Enchondromatosis, Ollier's disease, Skull base, Intracranial

Introduction

Enchondromas are common intraosseous benign cartilaginous tumours. When multiple, the condition called enchondromatosis, including Ollier disease and Maffucci syndrome (1), with a tendency to involve ends of long bones and flat bones, frequently presenting in the first decade of life (2,3).

Intracranial chondromas are very rare tumours (4,5), most frequent at the skull base with a predilection for the sphenoidal region (6) and less commonly involving the dura mater, choroid plexus, leptomeninges and brain parenchyma (7). They occasionally have been reported to arise at the sites of previous trauma (8).

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Case Report

A 25-year-old female patient, with known case of Ollier's syndrome of a history of multiple asymmetrical enchondromas (Fig. 1) associated with pelvic, tibial and femoral fractures during a period of 20 years, was admitted to our hospital with the chief complaint of right eyelid ptosis, progressive visual disturbance and bitemporal hemianopia, associated with marked deformation of the upper and lower extremities. She denied family history of similar disorder. She was alert and did not have history of headache, seizure attack or unconsciousness. No evidence of hemangioma was identified on detailed physical examination. All laboratory data were within normal limits.

Brain MRI revealed a lobulated heterogeneous mass measuring 34x26x23mm, located at pre-pontine cistern extending to suprasellar, right parasellar and right cerebellopontine angle and compressing the right optic nerve. Another small mass with similar characteristics was noted in left cerebellopontine angle. The masses showed low intensity signals on T1 (Fig. 2A), foci of hyperintensity on T2 (Fig. 2B) and contrast enhancement with a hypodense central core (Fig. 3). She then underwent trans-sphenoidal tumour excision.

Microscopic examination of the resection specimen showed multiple well delineated cartilaginous lobules without myxoid change separated by normal marrow fat and host lamellar bone (Fig. 4). Most parts of the tumor composed of evenly distributed chondrocytes with bland looking features devoid of mitoses. Another notable finding was focally increased cellularity accompanied with mild cellular atypia (Figs. 5A&B). No entrapment of the host lamellar bone in the neoplasm was identified on serial sections. With regard to the radiologic, clinical and histopathologic findings, the case was diagnosed as skull base enchondroma.



Fig.1: Plain radiograph of both hands revealing multiple enchondromas with calcification

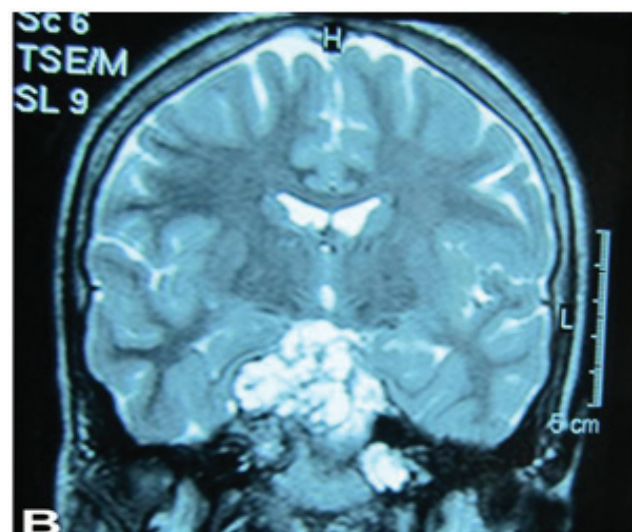
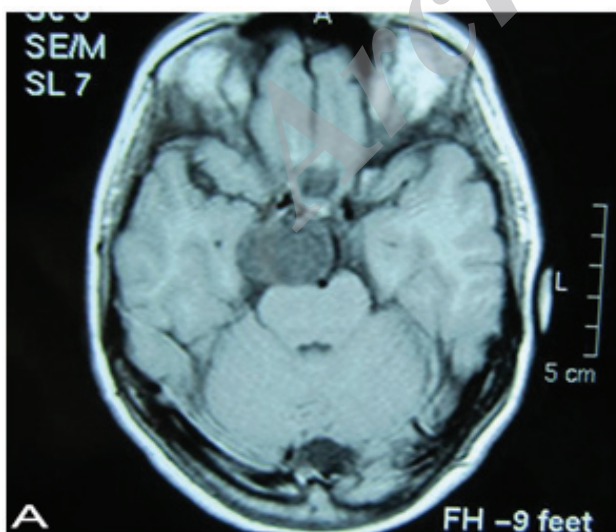


Fig.2: Brain MRI depicting lobulated intracranial mass in pre-pontine cistern involving the clivus, diagnosed as enchondroma, hypointense on T-1 weighted images (A) and depicting foci of hyperintensity on T-2 weighted images (B)

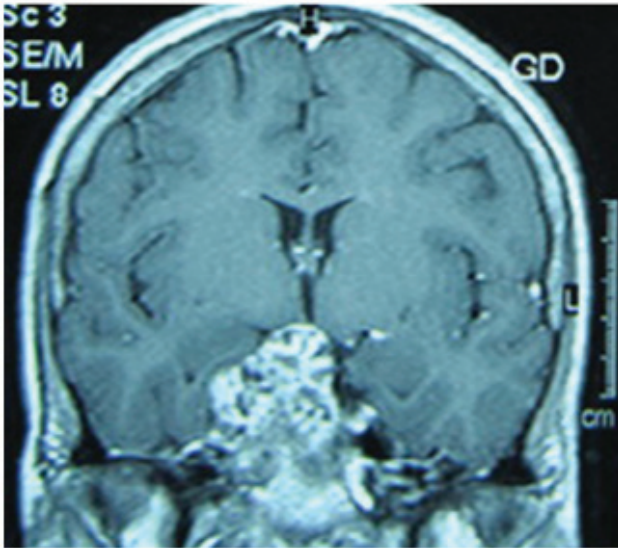


Fig.3: Coronal brain MRI showing marked contrast enhancement of Tumor

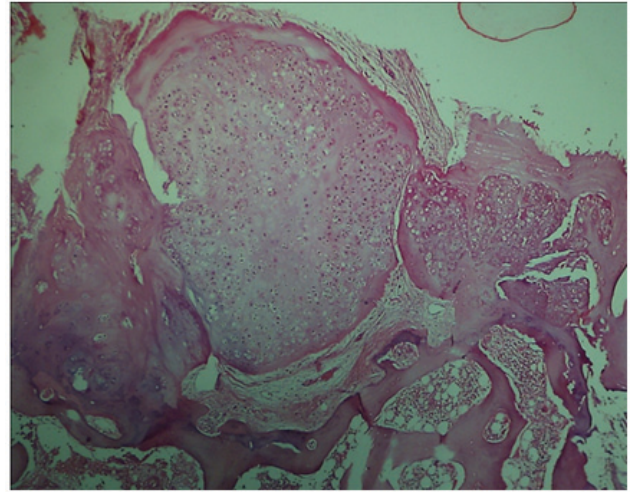


Fig. 4: Cartilaginous neoplasm consistent with enchondroma. Note well-demarcation and absence of bone invasion (40X)

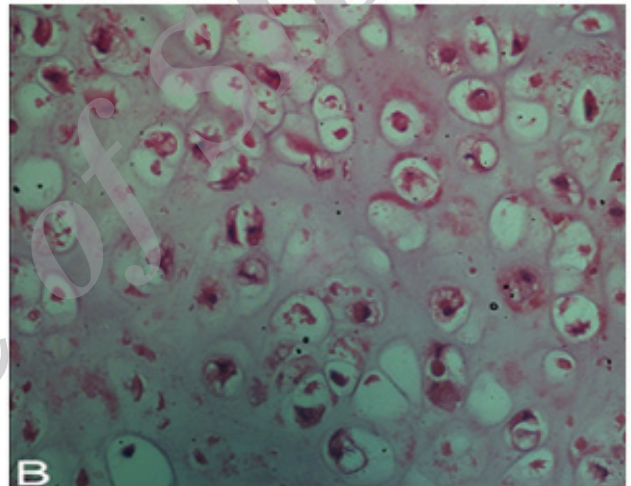
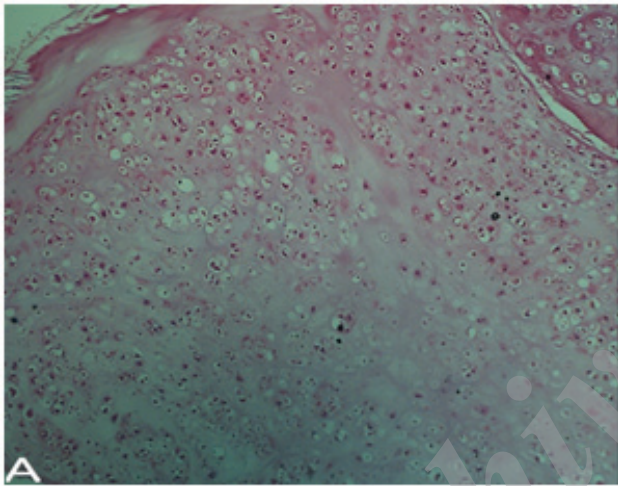


Fig.5: Focal hypercellularity and mild nuclear atypia of enchondroma (100×, A) & (400×, B). These findings are common in Ollier's syndrome and have no significance

Discussion

Intracranial chondromas are benign rare cartilaginous tumors accounting for about 0.2% to 0.3% of all intracranial tumors (4, 5). They may develop at any age, most frequently observed in the third decade (9). Despite of purported lack of sex predilection, a slight female predominance is evident in several case reports (10). The first reported case of intracranial chondroma was described by Hirschfield in 1851 (11).

These neoplasms are usually solitary, but they have also been reported as a component of Ollier's multiple chondromatosis (12) and

Maffucci syndrome (13).

Intracranial chondromas mainly involve the skull base, with special tendency to the sellar and parasellar regions (14-16). Dura, brain parenchyma and intraventricular space (8, 9, 17) are other reported sites of origin.

Various origins have been proposed for intracranial chondromas, the most common one being embryonic remnants of chondrogenic cells along the synchondrosis of the skull base (18). The chondromas arising from the dura matter, choroid plexus, and cerebral cortex have been assumed to arise from metaplasia of meningeal and perivascular fibroblasts (19). Proliferation of

ectopic embryologic rests of cartilaginous cells, traumatic displacement of cartilaginous elements or stimulation of fibroblasts to cartilaginous metaplasia by inflammatory processes are other proposed theories for the development of intracranial chondromas (18).

The slow growth of these tumors result in delayed clinical presentation, late diagnosis and large tumor size at the time of detection. Clinical manifestations are nonspecific, usually depend on tumor location and include headaches (8), proptosis, diplopia and varying degrees of visual acuity impairment as a result of orbital extension (15) forgetfulness and lack of concentration (18), Generalized tonic-clonic seizures (4), neurological deficits such as ptosis and bilateral hemianopia, increased intracranial pressure and psychologic/personality disorders. As we discussed above, our case had visual acuity impairment, bitemporal hemianopia and eyelid ptosis which progressed slowly.

Radiologic manifestations of the tumor are nonspecific and despite of developments, the definite diagnosis is histopathologic.

The histopathology of the biopsy samples of multiple enchondromatosis cases usually shows hypercellularity, double nucleated cells and nuclear hyperchromasia, suggesting a diagnosis of low-grade chondrosarcoma that should be ignored in the absence of clinical/radiologic features of chondrosarcoma (2). In current case, focal tumoral hypercellularity and mild nuclear atypia was ignored due to patient proved Ollier's disease as well as correlation with other histological and radiological data.

Radiologically, the tumor has been classified into two types: Type 1, which is the classic and more common type with homogenous and isodense pattern or mixed density with minimal to moderate enhancements and type 2 with hypodense cystic central area on CT scan (20). The most common differential diagnosis is meningioma, which is almost indistinguishable by radiology. Tanohata et al. reported two skull

base chondromas that exhibited delayed contrast enhancement on CT after administration of a high-dose of contrast medium. They suggested this CT feature to be of help in the differentiation of intracranial chondromas from meningiomas and neurinomas (21). MRI shows a well-circumscribed lesion without accompanying tissue edema that exhibits heterogeneous signal with intermediate to low intensity on T1-weighted images and mixed intensity on T2-weighted images (5).

The main complication of enchondromatosis is the risk of transformation to chondrosarcoma (22, 23) with an estimated risk of 20 to 50% (22). Histopathologic diagnosis of transformed chondrosarcoma may be difficult and in these instances, clinicoradiologic correlation is recommended (23). Any cortical bone destruction or extension into the Haversian systems or soft tissue favours chondrosarcoma.

In addition to the risk of developing chondrosarcoma, Ollier patients also have an increased risk of non-skeletal malignancies, especially glial tumours (24, 25).

Treatment of intracranial enchondroma is complete surgical removal, and the long-term prognosis is good. Local invasion or recurrence should raise the possibility of transformation to chondrosarcoma (9, 26).

Conclusion

Concluding from the above discussion, intracranial chondromas are well-differentiated cartilaginous neoplasms with a small proclivity for sarcomatous transformation.

Our case was valued because of its rarity, confusing and misleading histopathologic features and presentation in the background of a rare syndrome (Ollier syndrome); and indicates that histopathologic features of a bone neoplasm may be misleading if not accompanied by clinical and radiologic findings.

Conflict of interest

All authors declare there is no conflict of interests.

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