CASE REPORT

CHONDROMYXOID FIBROMA OF MASTOID PROCESS – AN UNUSUAL PRESENTATION OF A RARE TUMOR

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ABSTRACT: Chondromyxoid fibroma, a rare primary benign bone tumor usually affecting long bones of lower extremities in younger age group. An old patient presented with recurrent chondromyxoid fibroma of mastoid process of temporal bone, which is extremely rare. Chondromyxoid fibroma is known for recurrence after curettage, which often poses diagnostic difficulties and sometimes difficult to differentiate from more aggressive tumors like Chordoma and Chondrosarcoma. Though rare, Chondromyxoid fibroma should be considered in the differential diagnosis of primary skull base tumors.

KEY WORDS: Chondromyxoid fibroma; Primary bone tumors; Skull base tumors

INTRODUCTION: Chondromyxoid fibroma is a rare benign primary tumor which usually affects the metaphyses of the long bones of the lower extremities in childhood and young adults and constitutes less than 1% of all primary bone tumors. It is composed of chondroid, myxoid and fibrous tissue growth in a lobular pattern, infrequently with calcifications.1

We are presenting a case of this rare tumor with unusual presentation which carries significant clinical importance.

CASE REPORT: An 80 year old female patient came with complaints of foul smelling, purulent discharge of right ear associated with pain and right sided facial nerve palsy. The patient had undergone antral curettage three months ago for similar complaints and was diagnosed of chondromyxoid fibroma. On examination, foul smelling ear discharge and swelling over right mastoid was present. X-ray showed an indistinct expansile lytic lesion of right mastoid process. The patient underwent modified radical mastoidectomy. The histopathological examination of the specimen confirmed recurrence of chondromyxoid fibroma. She was a known case of carcinoma lip and had undergone radiotherapy six months ago.

Grossly, antral curettage specimen consists of multiple grey white, firm, irregular tissue fragments altogether measuring 3x3cm. Modified radical mastoidectomy specimen consists of multiple grey white, firm, irregular tissue masses largest ms.5x4x2cm. Cut section showed grey white homogenous areas with translucent cartilaginous areas [Fig.1].

Microscopically, the tumor showed lobules of chondroid matrix with myxoid material and areas of fibrous tissue [Fig.2]. Similar histological features were seen in the initial tumor and the recurrent tumor.

DISCUSSION: Chondromyxoid fibroma (CMF) is an uncommon benign primary tumor of bone that was first described by Jaffe and Lichtenstein in 1948.1 It occurs most frequently in young adults and usually develops in the metaphyses of the long bones of the lower extremities, followed by the flat
bones and the bones of the hands and feet. Craniofacial involvement is relatively rare. It manifests most frequently in the second and third decades of life, more often in males than in females. CMF is one of the least common tumors of bone, composing less than 1% of bone tumors and less than 2% of benign bone tumors.²

This case was an elderly female with CMF of mastoid process of temporal bone with recurrence and presented with aural fullness caused by otitis media with effusion and facial palsy of the same side.

The patient was on radiotherapy for squamous cell carcinoma of lip. There are incidences of bone tumors like osteosarcoma arising following radiotherapy,³ but we have not come across cases of chondromyxoid fibroma arising following radiotherapy in the literature.

The tumour has a tendency of local destruction and expansion towards the local tissues.⁴ The biological behaviour of this tumour has not been well studied, primarily because of the limited number of reported cases and the short duration of follow-up.⁵

In the present case, the tumor was arising from the mastoid process and expanded the mastoid facial nerve canal and leading to progressive facial nerve palsy.

Radiological signs of this tumor are the presence of a lobular, eccentric, single, lytic lesion, with expansion of the affected bone.⁴

Grossly, the tumors obtained by curettage contain numerous irregular fragments of tissue. Those located in the extremities are well limited in the outer surface and covered by a thin shell of newly formed periosteal bone or directly by the periosteum. Cut surface, shows a solid tumor mass of greyish-white or bluish grey colour, somewhat translucent, resembling cartilage. Only few cases show the frankly mucoid aspect with formation of cavities of different size produced by cystic softening of the mucoid tissue.

Microscopically, tumour is composed of lobulated or pseudo lobulated areas of spindle shaped or stellate cells without distinct cytoplasmic border and abundant myxoid or sometimes chondroid intercellular material separated by bands of more cellular tissue.⁶

A higher incidence of recurrence for CMF has been reported in the literature appears to be the consequence of the preference for curettage as the initial method of treatment, resection being performed only as a second surgical procedure,⁶ as it happened in the present case.

Chondromyxoid fibroma of skull base is a rare entity. Involvement of the temporal bone is particularly rare.⁷ It is a well-known tumor for its tendency for regional expansion and usual recurrences after the curettage of the lesion. Sometimes it is difficult to differentiate it from other more aggressive lesions, such as chordoma and chondrosarcoma histologically.⁸ The proper histological identification of the tumor and treatment may decrease the possibility of relapse. In conclusion, chondromyxoid fibroma should be considered as a rare benign tumor in the differential diagnosis of primary skull base bone tumors.

REFERENCES:

Figure 2. Chondromyxoid matrix with areas of fibrous tissue. [Haematoxylin & Eosin, x400]

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