Primary Chondroid Chordoma arising from the Petrous Temporal Bone: A Case Report

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Case Report

A 43-year-old female presented with a thirty month history of continuous headache and progressive difficulty in hearing, and with a two month history of progressive dysphagia, dysarthria, and left sided facial weakness. Neurologic examination revealed a group of left sided cranial nerve palsies involving the VIIth, VIIIth, IXth, Xth, XIth, and XIIth cranial nerves.

The plain films of the skull revealed irregular destruction with subtle calcifications along the superomedial aspect of the left petrous temporal bone. Routine axial brain computed tomography (CT) revealed a 4 x 4 cm sized hypodense mass with peripheral enhancement and internal calcified foci at the left cerebellopontine angle (CPA) region (Fig. 1). On coronal CT, the mass was downward extended through the skull base into the left parapharyngeal space. The lower extent of the growth was at the level of the soft palate (Fig. 2). On high-resolution temporal bone CT, the petrous portion and a part of the mastoid portion of the left temporal bone were destroyed. Superior and posterior walls of the left internal auditory canal (IAC) were also destroyed, but there was no actual widening of the IAC (Fig. 3).

A suboccipital craniectomy was done, and left
Fig. 2. a. Coronal CT shows mass extension through the skull base into the left parapharyngeal space. Intraluesional calcifications are also seen on the parapharyngeal area as well as on the CPA region.
b. On axial neck CT, the mass is well demonstrated. The left styloid process is laterally displaced.

cerebellar hemisphere was medially tracted. Then the part of dark-reddish tumor mass was exposed. It was surrounded by cerebellar dura mater and had a mixed consistency composed of gelatinous and fragile components. It was partially removed.

Microscopic examination showed that the tumor was composed of two areas with more cellular and less cellular components. The more cellular area was composed of monotonous physaliferous cells which had abundant vacuolated cytoplasm. A bluish chondroid matrix with several chondrocyte-like cells was also demonstrated in the cell sparse region (Fig. 4). There was no atypical spindle cell. This appearance was compatible with chondroid chordoma.

The patient was given external radiotherapy at a dose of 5000 rads over a period of five weeks. She is being on follow-up.

Discussion

Chordomas are neoplasms of notochordal origin which represent about 0.1 to 0.2% of all intracranial neoplasms (9). The notochord extends from the coccyx to the basisphenoid and has the role in formation of the axial skeleton. So, most of chordomas have predilection sites along the midline axis, such as sacrococcygeal region and clivus. During the fifth week of fetal life, in the region of the skull base, the notochord persues a sigmoid shaped path. Throughout its course, the notochord gives off

Fig. 1. Precontrast (a) and postcontrast (b) brain CT scans: A 4 x 4 cm sized left CPA mass shows internal calcifications and peripheral enhancement. Mass effect on the fourth ventricle is also seen.
Fig. 3. High resolution temporal bone CT: a. Axial scan shows destructive change involving the petrous portion and a part of the mastoid portion of the left temporal bone. Note the extensive destruction of the left jugular fossa. b. Coronal scan shows destruction of the superior wall of the left IAC without definite widening.

numerous dendrite-like branches, some of which penetrate the base of the skull anlage and others which ramify under the nasopharyngeal mucosa(10,11). Therefore, it is possible that portions of these dendritic processes of the notochord become detached during the development and afford a basis for the development of chordomas at unusual sites.

The term 'primary chordoma' has been coined for those chordomas which arise from unusual extraaxial sites by Shugar et al (10). The maxilla (10) and base of the temporal bone (7,8) have been described as such sites of origin in the literature.

The term 'chondroid chordoma' means a histologic variant of chordoma which shows cartilaginous tissues in addition to the classical picture of a chordoma. Embryologically, mesoderm surrounds the notochord and eventually produces a cartilaginous model of the spine. These mesodermal rests may become trapped in chordal vestiges and produce a cartilaginous component in the adult tumor. About 15% of all chordomas are chondroid chordomas. (2).

The case that we report is unusual in being both a primary as well as a chondroid chordoma. Our patient presented with left sided low cranial nerve palsies. The plain skull film and the CT findings were compatible with the radiologic characteristics of chordomas such as bony destruction and an extraosseous soft tissue mass with intrallesional calcifications (12). But, the site of the lesion was unusual for chordomas. With the location, other possibilities of schwannoma, glomus jugulare, metastasia, and cartilaginous tumor such as chondrosarcoma were also considered.

Cranial chordomas are usually diagnosed between the fourth and fifth decades of life. And there is no sexual predilection or a slight male predominance. But, chondroid chordomas present at an earlier age with female predilection. There is no definitely useful radiographic findings to differentiate chondroid chordomas from typical chordomas. The incidence of calcification appears to be higher with the former than with the latter. The recent report about MR fin-
ding of the chordoma suggested that chondroid chordomas had shorter T1 and T2 relaxation times than typical chordomas, perhaps because the watery and gelatinous matrix was replaced by the cartilaginous elements (13). The pathologic diagnosis of chordoma is based on the findings of large vacuolated ‘physaliferous cells’. Chondroid chordoma is characterized by areas resembling hyaline cartilage in addition to typical chordomatous areas.

The treatment of choice is total surgical removal, but cranial chordomas are seldom resectable because of the proximity of vital structures and the relative surgical inaccessibility of these areas (14). So, the treatment includes both surgical resection and radiotherapy. Postoperative radiotherapy may help to control the tumor over a longer period (3) and the response is dose dependent (15). In our patient, the surgical removal of the tumor was incomplete and followed by radiotherapy.

Average survival rates of chordomas vary from 4.2 years (16) to 7.7 years (4). Primary chordomas are known to have a better prognosis than typical chordomas in the usual sites (10). According to Heffelfinger et al. (2), chondroid chordomas also have a better prognosis with the mean survival of 15.6 years. Further follow-up is required for our patient to confirm this.

REFERENCES

12. Utne JR, Pugh DG. The roentgenologic aspects of chordoma. AJR 1955; 74:593-608