Clivus Chordoma with an arachnoid cyst, coincidence or causative factor?

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ABSTRACT

Clivus chordoma coincidence with arachnoid cyst is rare. We report a rare case of clivus chordoma associated with arachnoid cyst.

Keywords: Chordoma; Skull Base; Arachnoid Cyst; Clivus

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INTRODUCTION

The clivus is the surface of a portion of the occipital and sphenoid bones in the base of the skull [1]. It is surrounded by the neurovascular structures of the brainstem, as well as both internal carotid arteries. Tumors of the clivus can be benign (chordoma) or malignant (chondrosarcoma) [2].

Chordomas are rare, aggressive, slow-growing, invasive, and locally destructive tumors that arise from the notochordal, a structure that appears in embryonic stages and guides the growth of the bony skull and spine. Normally, notochord remnants form part of the intervertebral discs. A chordoma occurs when additional notochord cells are enclosed by the developing bones. Also these tumors are slow-growing and benign. They may invade nearby structures and destroy surrounding tissue and spread to other parts of the body [3]. In rare instance it could be associate with arachnoid cyst and the first symptoms may be caused by an arachnoid cyst [4,5].

Most arachnoid cysts are probably present at birth, or develop soon after. Once they are formed, they are thought to remain stable, apparently in some kind of equilibrium with the rest of the intracranial space [3].

CASE PRESENTATION

History and Examination

A 32-year-old man presented to our hospital with a 3-year history of headache. A magnetic resonance image (MRI) of the brain revealed an erosive large solid and compendious cystic mass, involving the clivus and a cystic mass in left temporal lobe (Figure 1). The patient was admitted to our hospital for further investigation. Neurological exam was normal except intermittent photophobia.

Computed tomography (CT) showed that the tumor destroyed the lower clivus. The tumor was localized in the extradural space without calcified components. There was also a cyst in left temporal lobe (Figure 2).

Surgery was performed by endoscopic transnasal trans-sphenoclival approach because tumor was mainly extended toward the posterior part.

Histopathological Findings

Three types of chordoma can be identified histologically: classic, chondroid, and dedifferentiated. The classic microscopic appearance of a chordoma is a lobulated growth of cords and islands of polygonal tumor cells suspended in a myxoid-mucous background. The epithelioid cells are slightly elongated, with associated large mucus-containing physaliphorous cells [6] (Figure 3). The nuclei are round and uniform, although some exhibit considerable pleomorphism.

DISCUSSION

Chordomas are tumors arising from rest of primitive

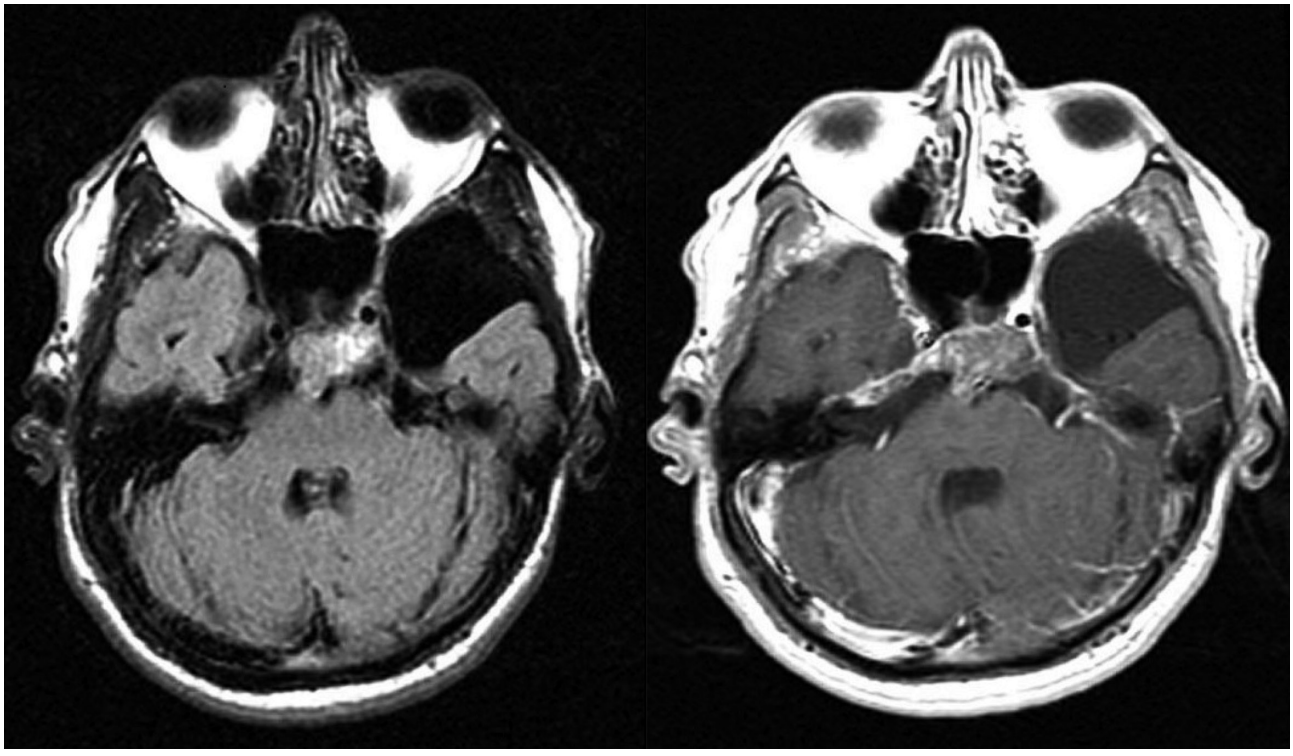

Figure 1. Axial preoperative MRI without and with Gadolinium shows extension of the clivus chordoma (heterogeneous enhancement) with an arachnoid cyst in left temporal lobe.

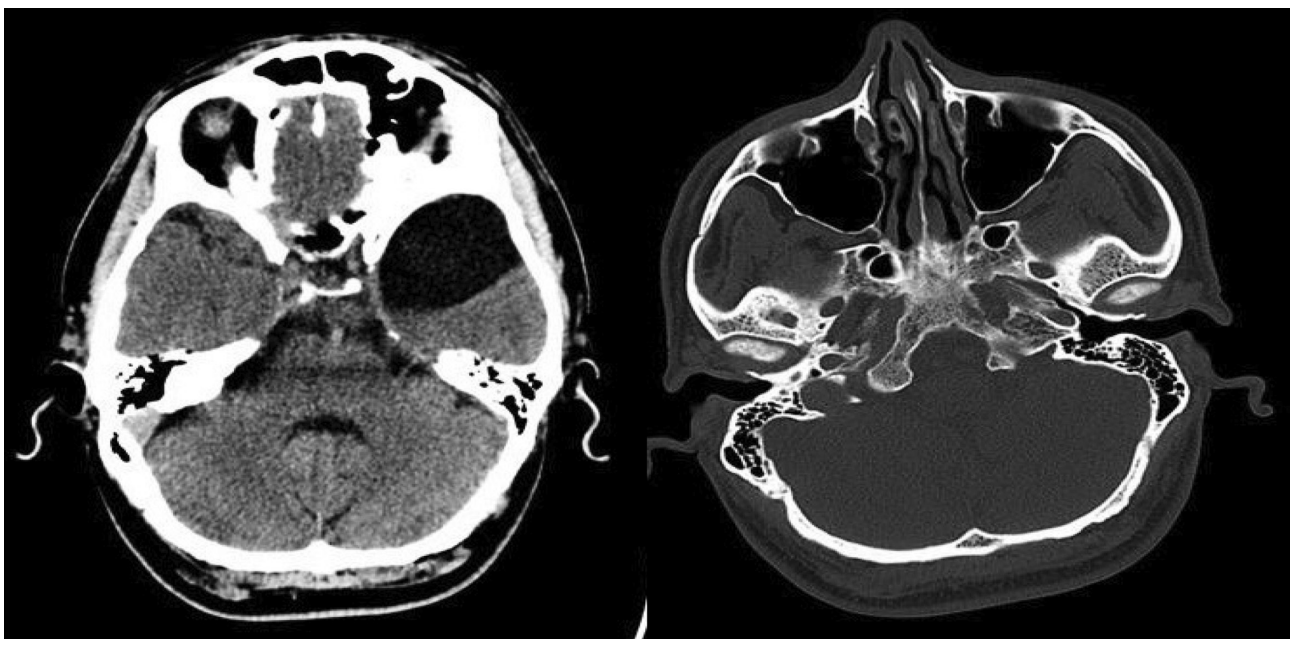

Figure 2. Axial computed tomography image shows the erosion of the clivus (right) and arachnoid cyst (left).

notochord along the craniospinal axis. The most common site is the sacrococcygeal region (50%), followed by the skull base (35%) and vertebral bodies (15%). Chordoma account for approximately 1% of all intracranial tumors and the most common site is retroclival region [7]. Chondroid chordoma, first described by Heffelfinger in 1973, is a variant of chordoma that straddles the bridge between conventional chordoma and chondrosarcoma. About 28-34% of skull base chordomas are of the chondroid variety [8]. Chordoma usually occurs between the fourth and fifth

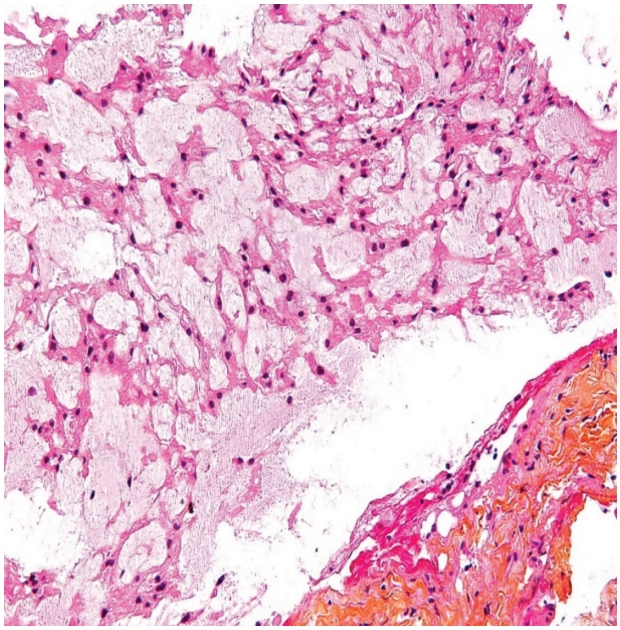

Figure 3. The tumor cells have abundant eosinophilic cytoplasm with moderate nuclear atypia and form anastomosing cords. Epithelioid appearance due to abundant eosinophilic cytoplasm.

decades of life, whereas chondroid chordoma present at an earlier age and has a female predilection [1,9]. Chondroid chordoma has a lower recurrence rate and better survival as compared to conventional chordoma [9,10]. However, the location is challenging to operate, it commonly occur the clival region. Safe, sufficient decompression is the most important issue for lower clival tumors, particularly in cases involving the foramen magnum. Traditionally transcranial transnasal, extended frontal, trans-maxillary, and transoral approaches are performed for anterior approaches to the foramen magnum [12,14,15]. However, such approaches carry a high risk of morbidity involving neurovascular structures. These traditional approaches reportedly offer a total resection rate of 44-83%, neurological morbidity in 0-80% of cases, vascular injury in 9-12%, and cerebrospinal fluid (CSF) leakage in 8.3-30% [11,13,16].

Arachnoid cysts most likely originate from a minor aberration in the development of the arachnoid that leads to splitting or duplication of the membrane [4,17,18]. It has also been postulated that the cyst develops from a defect in condensation of the mesenchyme or from abnormalities of CSF flow [18,10,20,21]. The association of other developmental abnormalities of the brain, such as heterotopias, lent support to this developmental theory [19]. In the 208 reported cases of arachnoid cysts analyzed by Rengachary and colleagues [19,20], they found that the structural features of the arachnoid cyst wall that distinguish it from the normal arachnoid membrane were as follows: 1) splitting of the arachnoid membrane at the margin of the cyst; 2) a thick layer of collagen in the cyst wall; 3) the absence of traversing trabecular processes within the cyst; and 4) the presence of hyperplastic arachnoid cells in the cyst wall. In a study of five cases, Rengachary et al reported that the structure of the arachnoid cyst wall was similar to that of the normal arachnoid membrane and that the inner surface of the arachnoid cyst wall was formed of one or several layers of arachnoid cells with slender processes, which contained large extracellular spaces but no microvilli [19]. These cysts appeared to be truly intra-arachnoid in location and to be formed by splitting or duplication of the arachnoid membrane [19,20]. According to the ball-valve hypothesis, an anatomical fissure of the cyst wall functionally acts as a one-way valve, allowing free entrance of CSF but preventing its exit into the subarachnoid space. One-way pulsatile movement of CSF has been demonstrated by cine-mode magnetic resonance imaging studies and confirmed at endoscopic operations by inspecting the inside of the cyst wall [20]. The theory of an osmotic gradient between cystic contents and CSF lacks support given that the cystic content is similar in composition to CSF [19].

In patients with tumor-associated cysts, growth of the tumor or secretion of growth factors can have a mechanical or chemical effect on the arachnoid [20].

Previous studies assume that chordomas displace surrounding soft tissues without invasion [21] research suggests that chordomas either invade the bone or only compress the adjacent arteries and nervous structures without invasion, emphasizing the fact that the tumor invades the submucosal layer and not the dura and vital neurovascular structures, even in the advanced stage [21-23] and possibility an unusual temporal cavity (arachnoid cyst) was seen.

There are few reports about the clivus chordoma and arachnoid cyst. Herold et al reported Clivus Chordoma in Continuity with a Large Pontine Cyst [23]. In their case the cavity was not a separate arachnoid cyst. It was in continuity with the solid part of the neoplasm. Doglietto et al also reported a case of clivus chordoma associated with a pontine arachnoid cyst, he postulated that the tumor creates a local flow alteration of cerebrospinal fluid leading to the formation of the cystic lesion [24].

CONCLUSION

There are few reports about the clivus chordoma

associate with arachnoid cyst. Whether this is a coincidence or causative factor, further studies is needed to answer this question.

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