Diagnostic Clues and Treatment of Intradural Cranial Chordoma

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To the Editor: A 69-year-old man was admitted to our center for poor appetite and malaise for 10 days. Physical examination revealed impaired consciousness and dementia appearance. The head computed tomography (CT) showed a suprasellar lesion of mixed densities without bone invasion, as well as the eggshell calcification presented as interrupted little patches of high-density encircling focus [Figure 1a]. The magnetic resonance imaging (MRI) showed an approximately 38.8 mm × 33.1 mm × 36.4 mm lesion in the suprasellar region with an unclear contour and an ampliative third ventricle which was caused by the tumor compression and invasion [Figure 1b-1e]. The suprasellar lesion presented isointense on T1-weighted image (WI) [Figure 1b] and hyperintense on T2-WI [Figure 1c]. The postcontrast lesion was obviously enhanced with cellular nonenhanced region concentrating on center of the tumor [Figure 1d and 1e]. It is clear that a lobulated lesion [Figure 1b-1d] along with a honeycomb-like manifestation [Figure 1d and 1e] could be detected in figures. No obvious abnormalities were identified in serological tests of pituitary hormones, adrenocortical hormones, thyroxines, and sex hormones.

Eventually, the patient received the diagnosis of suprasellar tumor inclined to craniopharyngioma before operation. Therefore, a frontobasal lateral surgical approach for resection was performed. The tumor, short in blood supply, was yellow-white, tough, and hard. The neoplasm was entangled with adjacent structures and grew into the retrochiasmal area. The intraoperative frozen pathology revealed a tumor of eosinophil. Given the underlying complications might occur after resecting the adjacent tough tissues closely adhering the infrachiasmatic lesion, we only performed an incomplete resection, in which approximately 50% of the tumor was resected. Seven days later, the routine pathology affirmed that the morphological appearance was in accordance with the chordoma [Figure 1f]. In the meanwhile, the immunohistochemistry showed positive reactions of S-100, CK (AE1/AE3), CK5/6, vimentin. On the discharge day, the patient was in a light coma and recommended for a rehabilitation therapy. Followed up for seven months, the patient had gradually regained consciousness but was presented with uracratia and recent amnesia.

Chordomas are rare malignant neoplasms that arise from persistent notochord elements.[1] Approximately, half of all chordomas arise in the sacrococcygeal area, 30% in the base of the skull, and 20% in the mobile spine. Intradural cranial chordomas (ICCs) are extremely rare cases that only 45 cases have been reported to the best of our knowledge. Given the limited data of ICCs, we report a case of intradural and extraosseous chordoma located in the suprasellar region and review the relevant literatures. The median age at the time of diagnosis for chordomas is 60 years, with skull base presentations commonly affecting a younger population including children, and they are more common among males than females. After reviewing literatures,[2-4] we found no obvious difference in the sex ratio between chordomas and ICCs. Typical chordomas occur along the neuraxis, especially at the cranial and caudal ends. The occurrence rate of ICCs accounts for approximately 4% of all the skull base chordomas.[2,4] They mostly locate in the prepontine region, rarely in the intrasellar, suprasellar, pterional region, pineal, dorsum sellae, Meckel's cave, cerebellum, hypothalamus, tentorium cerebella, and foramen magnum. Occasionally, intracranial or craniospinal disseminations can occur. It is, sometimes, also difficult to differentiate chordomas from meningiomas, craniopharyngiomas, neuroepithelial cysts, schwannomas, arachnoid cysts, and gliomas.

CT with bone window is highly efficient in detecting bone destruction, calcification, and bleeding, which can be frequently seen in chordomas. MRI has also been recommended for the primary tumor assessment because of its better display in multiplanar views and delineation of different soft tissue components of tumors and their surrounding structures. Commonly, chordomas appear iso/hypointense on T1-WI and hyperintense on T2-WI with heterogeneous enhancements of extraskeletal components. A characteristic MRI finding of chordomas is the lobulated appearance.[5] And the septations, which make tumors lobulated or latticed, are enhanced.[5] In addition, the alveolate presentation is also a significant finding in most cases of reported ICCs. Therefore, we speculate that lobulated and alveolate appearances, as well as characteristic MRI images, are useful diagnostic clues of ICCs. In our case, the suprasellar lesion showed isointense on T1-WI and hyperintense on T2-WI with heterogeneous enhancements. A typical lobulated and alveolate presentation was also available. Although this time we misdiagnosed the case with these accessible

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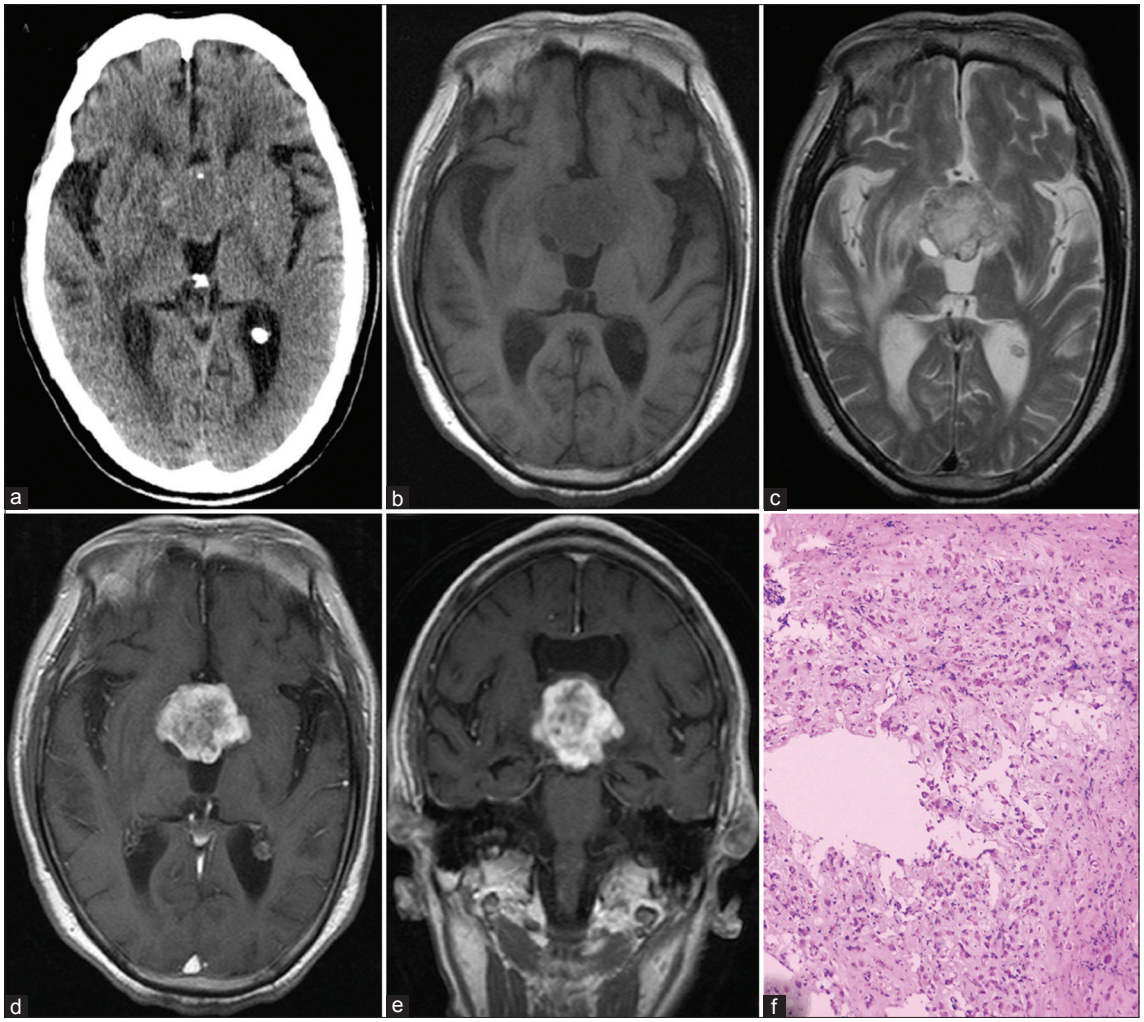

Figure 1: Head computed tomography, magnetic resonance imaging, and pathological picture of our case. (a) The cross-sectional view of computed tomography. (b) Cross-sectional view of T1-weighted image. (c) Cross-sectional view of T2-weighted image. (d) Enhanced magnetic resonance imaging, the cross-sectional view. (e) Enhanced magnetic resonance imaging, the coronal scan. (f) Histology hints physaliferous cells and mucin formation with the diagnosis of chordoma (H and E, original magnification ×160).

but unrealized diagnostic clues, we believe that, with reminders from these possible diagnostic clues, we could avoid similar faults in similar cases in future.

The treatment for classic intracranial chordoma is complete resection when possible, and there is no clear evidence that the radiation therapy contributes to a good outcome. On account of limited data of the reported cases, there is no exact consensus whether radiation therapy is effective in ICCs. And in our case, no radiation therapy was performed and surely our short time of follow-up and individual experience cannot deny the effect of the radiation therapy. The deficiency of clinical data of ICCs makes diagnosis and treatment difficult, and a relatively large independent research only contained six cases.[4] Therefore, more studies of ICCs are needed to help diagnosis and improve treatment.

In brief, the common location of ICCs is the prepontine region. The ectopia ICCs call for differential diagnoses. Both CT with bone window and MRI are beneficial tools in the diagnosis of ICCs. Possible diagnostic clues including characteristic images and lobulated and alveolate appearances can conduce to a better preoperative diagnosis. Complete resection is recommended when possible, and currently there is no evidence to determine whether radiation therapy is effective in treating ICCs.

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Conflicts of interest
There are no conflicts of interest.

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