Clival Cystic Chordoma in Children with Confused Magnetic Resonance Imaging

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There are some kinds of neoplasms to be differentiated in the clival area, such as chordoma, invasive pituitary adenoma, craniopharyngioma, trigeminal neuroma, epidermoid cyst, and epidermoid tumor. A 12-year-old girl had admitted with a chief complaint of diplopia. On computed tomography and magnetic resonance(MR) image, cystic mass of about 4cm in diameter which partially destroyed bony structure at posterior part of the clivus was compressing brainstem slightly at anterior direction. This mass showed hyperintensity on T2-weighted MR image, hypointensity on T1-weighted MR image and especially hyperintensity on diffusion image, but no contrast enhancement. So, We suspected an epidermoid cyst and planned biopsy through a transsphenoidal approach. Fortunately, tumor was removed completely. This tumor was considered as epidermoid cyst on MR image. But, it was confirmed as chordoma in pathologic diagnosis. Therefore, we report this case and it may be helpful to make a differential diagnosis in clival cystic chordoma preoperatively that has multilobulation, bony erosion, and calcification on radiologic findings.

KEY WORDS: Clivus · Cyst · Chordoma · Childhood · Transsphenoidal approach.

Introduction

Most of the clival chordomas are frequently noted among adults in their 40’s and 50’s and are very rare among childhood. Only 75 cases have been reported for intracranial chordoma in childhood in 19872,6), but exact count was unknown so far. Furthermore, the radiologic finding rarely shows pure cystic components. Chordoma, a tumor deriving its origin from notochordal remnant and developing along craniovertebral axis, is classified clinically as a malignant tumor because of its invasive nature into adjacent structures and its wide-spread metastasis even if it is classified histologically as benign tumor because of its slow growth rate4).

In this report, we removed a cystic clival tumor completely in a 12-year-old girl via transsphenoidal approach which was initially suspected of epidermoid cyst by magnetic resonance(MR) image especially on diffusion weighted image but confirmed as chordoma on histologic examination. Hence, we expect our case would be helpful in differentiating clival tumor with cystic nature in radiologic examination before surgery.

Case Report

A 12-year-old girl with a chief compliant of diplopia, which had been developed seriously 4 months before, visited our hospital. She didn't have any problems in her family and past history. In her neurologic examination, she had alert mental state and right sixth nerve palsy. Also, there was a slight increase in prolactin level.

There was bone destruction in computed tomography(CT) such as the posterior part of the clivus and central lesion, including dorsum sellae to the half of the upper clivus, which has about 4cm-long cystic mass and compresses brainstem posteriorly. MR image finding shows a relatively heterogeneous multilobulation with low signal intensity on T1-weighted image and homogeneous manifestation with high signal intensity on T2-weighted image, especially in a diffusion image (Fig. 1). In these situations, we strongly suspected an epidermoid cyst with the differential diagnosis of cystic chordoma, chondroid tumor, cystic ependymoma, and so on.

We had planned a biopsy through transsphenoidal approach for the definite histological diagnosis, and fortunately it was possible to remove a tumor completely by only removing a part of the clivus. The lesion was suspected of a cystic mass before the surgery, and showed the grayish gelatinous pattern without blood vessel distribution. In a histological finding, we could observe a lobulated region composed of fibrous septum and a cordlike-pattern inside...
myxoid stroma. And cytoplasmic vacuolation, which generates bubble-like shape called physaliphorous cell, was found and in an immunohistochemical finding, a positive reaction to epithelial membrane antigen (EMA), cytokeratin (CK), S100 protein, and vimentin and a negative reaction to GFAP were observed (Fig. 2). Therefore, it was confirmed as chordoma in pathologic diagnosis.

Her symptoms of diplopia and sixth nerve palsy improved after the surgery and serum prolactin level turned to a normalization. We could confirm that the tumor was completely removed through a MRI (Fig. 3).

Discussion

Intracranial chordoma accounts for 0.2% of all intracranial tumors and typically is central lesion ranging from the clivus to the sellar turcica. This kind of tumor mostly comes up among people in their 40’s and 50’s, and very rare in infancy and childhood, and their sex ratio is 2:1 (male:female). Chordoma is located in the sacrococcygeal (49~54%), vertebral (20~24%) or intracranial (26~36%) areas. It is known that although sacrococcygeal region is the most common part of chordoma in adults, a tumor within the base of skull is more common in childhood.

Histologically, these tumors show a slow growth rate and are benign. However, it is classified as malignant clinically because of its critical locations, locally aggressive nature, high rates of recurrence, and tendency to metastasize.

Pathologically, the gross finding of chordoma is grayish gelatinous semitranslucent and its histological findings lie on the facts that it contains lobules composed of physaliphorous cells made by fibrous septa and shows a hyperchromatism or pleomorphism in nucleus. The immunohistochemical profile of reactivity with antibodies to vimentin, CK, EMA and S100 protein generally distinguished chordoma from sarcomatoid, round cell, or myxoid neoplasms.

Generally, following factors in radiological findings are observed for suggesting chordoma as low or isosignal intensity by CT, heterogeneous contrast enhancement by enhanced image and bony erosion as well as bone destruction accompanying calcification or bone fragments. A heterogeneous manifestation that shows a region divided by septum with low signal or isosignal intensity on T1-weighted image of MR image, a homogeneous manifestation with high signal intensity on T2-weighted image, and over 80% of heterogeneous contrast enhancement were observed. Differential diagnosis include...
invasive pituitary adenoma, craniopharyngioma, trigeminal neuroma, dermoid and epidermoid cysts, and so on. In this case, epidermoid cyst was strongly suspected after considering the findings including low signal intensity with rather heterogeneity on T1-weighted image, high signal intensity with homogeneity on T2-weighted image, especially in diffusion, and no contrast enhancement by enhanced image. But even if there was no contrast enhancement by enhanced image, it was possible to differentiate chordoma from epidermoid cyst for the following factors of the tumor such as its location, the degree of invasion to adjacent structures, and consisting of many lobules.

The choice of surgical approach in treating clival chordoma depends on many factors including the location of tumor, patient's clinical status, previous surgery and/or radiation, and the experience and philosophy of the surgeon. Transsphenoidal approach is primarily preferred for removing chordoma located on the upper and middle part of the clivus. Transsphenoidal approach provides excellent exposure to the sphenoid sinus, sellar turcica, and upper and middle part of the clivus. But its disadvantages include limited lateral and inferior exposure, a deep and narrow field, and lack of proximal control of the cavernous carotid artery. However, in this case, a tumor, showing cystic nature and whose lateral extension is not severe, can be removed completely through a transsphenoidal approach within a limited exposure.

Conclusion

Clival cystic chordoma in childhood was very rare and was difficult to differentiate from epidermoid cyst through clinical and radiological findings. However, it was possible to differentiate clival cystic chordoma from epidermoid cyst for the following respects: clival cystic chordoma, unlike an epidermoid cyst, shows a multilobulation caused by fibrous septa in addition to the location, and is epidermal tumor invading adjacent structures showing a bone destruction. We have come to conclude that this pattern of cystic tumor can be removed safely within a limited exposure by the transtuberculum approach while minimizing its surgical morbidity.

References