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ABSTRACT

We report an extremely rare case of an endodermal cyst of the cranio-cervical junction located dorsally to the brainstem and upper cervical spine in a 27-year-old female presented with occipital headache, vertigo and pain in both shoulders. Neurological examination showed neck stiffness with bilateral XIth nerve palsy. Magnetic resonance imaging revealed a cystic lesion at the cranio-cervical junction and slight compression of the brain stem. The lesion was totally removed through the posterior approach. The histological diagnosis was endodermal cyst. To our knowledge, the only one such case has been reported in the literature.

INTRODUCTION

Endodermal cysts can be found in literature by a variety of different names including neurenteric, epithelial, bronchogenic, enterogenous, respiratory and foregut cysts. These cysts are a benign congenital condition resulting from the persistence of an abnormal communication between endoderm and neuroectoderm at 3 weeks life of the embryo. This malformation is encircled by a mucosal secreting epithelioma mimicking the normal gastrointestinal epithelioma tractus (1). Exact histopathological diagnosis may be hard to establish, since there are many similarities with other cystic lesions such as Rathke cysts, colloidal cysts, and/or cystic teratomas (9). Intracranial locations are rare, and location of such cysts in the cranio-cervical junction is exceptional. They are located in the midline, in ventral or ventrolateral locations. Endodermal cysts may occur at any age, but there is a slight predominance of male patients in their forties. Since they are slow growing tumours, many patients have only mild symptoms relative to the tumour size. The main symptoms are due to compression and mass

Keywords
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effect. However, cases of recurrent aseptic meningitis can be found in literature (2).

Neurenteric cysts are treated surgically. With only partial resection, endodermal cysts may recur at the original site on long-term follow-up (3-6). The incomplete resection may be associated with postoperative malignant transformation or widespread cranial-spinal dissemination. Almost all authors advocate the aggressive resection to reduce the possibility of endodermal cyst recurrence (7, 8).

CASE REPORT

We present a case of a 27-year-old woman admitted with a history of recurrent episodes of occipital headache, vertigo and pain in region of both shoulders of six months duration prior to admission. She had no difficulty with swallowing or speech, and the strength of all extremities, gait, and coordination were normal. The neurological examination presented only moderate neck stiffness with bilateral XIth nerve palsy. There was neither sensibility deficit nor extra neurologic signs.

Magnetic resonance imaging (MRI) of the cranio-cervical junction revealed a well-defined, round, intradural cystic lesion located dorsally to the spinal cord and the medulla, extending from the foramen magnum to the upper level of C1 lamina, with neither bony nor soft tissue associated abnormalities. T2-weighted images displayed a hypointense signal, and T1-weighted MRI demonstrated a hyperintense mass without enhancement. The lower medulla and C1 spinal cord were found to be slightly compressed in the posterior aspect in the sagittal image. This intradural extraaxial process was measuring 20,7 x 12,7 x 8,3 mm (Figure 1). Preoperative CT scan showed a high density area at the cranio-vertebral junction, well delimited, dorsal to the medulla and upper cervical spine. In our case the cyst appeared spontaneously hyperdense.

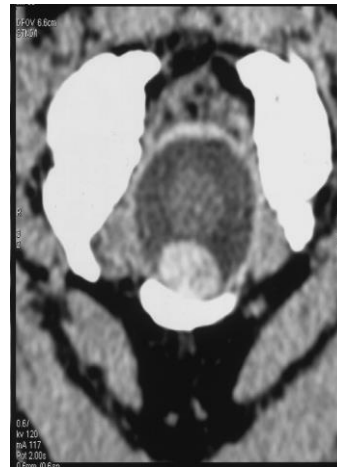


Figure 1. Cranio-cervical junction MRI showing intradural cystic lesion located dorsally to the spinal cord and the medulla.

The patient was operated in a prone position through a posterior approach and midline incision. Surgery consisted in a reduced sub-occipital craniotomy

associated to partial C1 laminectomy. After opening of the dura a thin-walled yellowish cyst was noted below atlanto-occipital membrane, in the region of cerebellomedullaris cistern. It was floating in liquor, attached with bulbo-medullary junction only with thin arachnoidal connection. The lesion had not adhered to any other surrounding structures. Surgical excision was complete in one piece.

On pathohistological examination, the cyst membrane was underlined by unistratified cylindrical enteroid cells with basal nucleus and apical muco-secreting pole, with supra-nuclear accumulation of alcain-blue positive mucine. The diagnosis was endodermal cyst.

Postoperatively, the patient showed no neurological deficits. In the next few months' clinical signs resolved completely. Postoperative CT (Figure 2) and MRI revealed no evidence of a residual cyst.

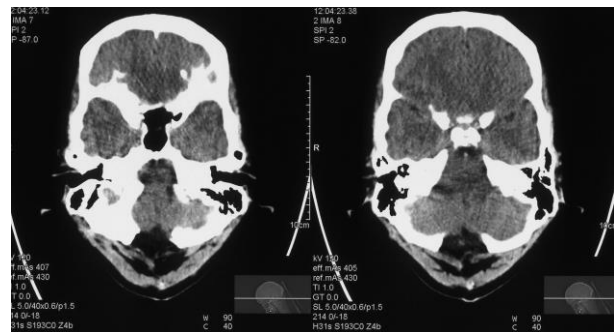


Figure 2. Postoperative CT scan showing complete resection of cyst.

DISCUSSION

Endodermal cysts are congenital abnormalities belonging to notochordodysraphies which are result of an abnormal adherence between ectoderm and

endoderm. A failure during embryogenic development is probably responsible for cyst formation (1,9,10,11). The pathogenesis of this lesion is not clarified. However, the most convincing hypothesis is a dysgenesis of the endoderm from the neuroectoderm in the 3rd week of foetal development (12). Usually, these lesions occur at the lower cervical and upper thoracic region, and their ventral origin is suggested by defects in the vertebral bodies. The lesions may be either intradural or extradural (13). The present case is unusual because cyst is found in the cranio-cervical junction and exceptionally unusual because of dorsal location, representing the only one such case reported in the literature (14, 15, 16). They show slow growing rate, remaining asymptomatic for a long time, but without surgical removal they can produce a brain stem and spinal cord compression syndrome followed-up by a neurological impairment. In this case, according to our opinion, surgery was justified in order to achieve total removal of the cyst and to prevent recurrence, malignant transformation or subarachnoid dissemination.

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