CASE REPORT

Skull base hydatid cyst with intracranial extension presenting as vocal cord palsy: a case report

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ABSTRACT. Hydatid disease of the skull base is extremely rare, and intracranial extension of hydatid cysts through the skull base is even rarer. We report an interesting case of a 42-year-old man who presented with features of right vocal cord palsy. The diagnosis of hydatid cyst was made based on his history and on pre-operative MRI and was confirmed by surgery and histopathological examination.

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Hydatid disease is caused by infestation of the larval stage of tapeworm of the genus *Echinococcus*. Multiple echinococcal species produce hydatid disease, the most common causative species being *Echinococcus granulosus*. *Echinococcus multilocularis* is less commonly described but is more invasive, mimicking a malignancy [1–3]. Liver and lung involvement are most common, whereas bone and soft tissue involvement are rare [1–5]. Only a few cases of skull base involvement have been reported in the literature [4, 6, 7]. Our case is an unusual location of hydatidosis (*i.e.* hydatid disease of the skull base with intracranial extension). Usually, hydatid cyst of the head and neck region presents with local swelling and non-specific symptoms. Our patient presented with progressive hoarseness of voice. Compression of the vagus nerve by an intracranial or extracranial hydatid cyst is rare [4].

Case report

A 42-year-old man presented to our ENT department with hoarseness of voice and incessant cough of six months' duration. He was referred to the neurosurgical unit for further management. He had undergone repeated surgeries for excision of hydatid cysts in the right posterior triangle of the neck on three occasions over the past 27 years. The last surgery consisted of aspiration of the cystic mass in the neck followed by injection of a sclerosant. On physical examination, fullness was noted in the right upper posterior triangle of the neck. Nasopharyngolaryngoscopy revealed decreased palatal movement, nasopharyngeal congestion and features of right vocal cord palsy. No abnormality except neutrophilia (83%) was found on laboratory studies.

CT was carried out to look for a compressive lesion along the vagus nerve or recurrent laryngeal nerve. CT revealed a large multiloculated, septated, cystic, thin peripherally enhancing mass in the right posterior skull base with destruction of the right occipital and adjacent part of mastoid bones. The extracranial component of the lesion extended to the right jugular foramen, inferiorly along the right paravertebral region up to the C3 vertebral level. There was intracranial extension into the posterior cranial fossa in the right cerebellopontine region (Figure 1). Atrophy of the right sternocleidomastoid and trapezius muscles was noted. MRI was performed before surgery to look for further details of the intracranial extension of the lesion. MRI revealed a large cerebrospinal fluid (CSF) signal multivesiculated, multiseptated lesion in the right posterior skull base giving a "bunch of grapes" appearance (Figure 2). Thin hypointense peripheral rim ("rim sign") and irregular linear low-signal-intensity structures within the cyst suggestive of collapsed membranes ("serpent sign") were noted (Figure 3). These were best seen on the heavily T_2 weighted sequence. The lesion extended intracranially through the destroyed occipital bone into the posterior fossa in an irregular pattern, to lie in the inferior aspect of the right cerebellopontine cistern. There was compression and displacement of the medulla, right lower cranial nerves and ipsilateral vertebral artery towards the contralateral side; the lesion also compressed the adjacent part of the right cerebellar hemisphere. It was unclear from the imaging whether the intracranial cysts were intradural or extradural in location. The extracranial component was in the right jugular foramen and suboccipital region along the carotid sheath. A pre-operative diagnosis of recurrent hydatid cyst was considered.

The patient underwent surgery with right lateral suboccipital craniectomy and total excision of the cysts. There was dense fibrosis in all the planes of the neck,

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Figure 1. (a) Axial and (b) coronal contrast-enhanced CT images show a complex solid and cystic mass at the right posterior base of the skull eroding the occipital bone (white arrows). There is a jugular fossa component with suboccipital and intracranial extension into the posterior fossa (black arrows).

possibly due to the repeated surgeries and the sclerosant injected earlier. Several cysts were extracted from the subcutaneous and muscular planes and from within the occipital bone. The right occipital bone was eroded and several cysts were also seen in the extradural plane along the posterior aspect of the right petrous bone. The intracranial cysts were entirely extradural in location. Intraoperative ultrasound was used to look for any cysts in the intradural compartment. The dura was not opened during the surgery and there was no CSF leak during the surgery. At the end of surgery, no obvious residual cysts were seen in any of the planes. This was confirmed using intraoperative ultrasound evaluation of the operative field. The post-operative period was uneventful. The hoarseness of the voice did not improve, and the patient did not develop any new neurological deficit. A post-operative CT scan did not reveal any obvious residual

cyst. Surgical pathology confirmed the diagnosis of hydatid cyst with amorphous lamellated material and daughter scolices. The patient was prescribed albendazole therapy in cycles of 4 weeks each with a break of 2 weeks between each cycle. This was done to prevent recurrence of the disease.

Discussion

Hydatid disease is a common parasitic infestation with varying imaging appearances; it can involve any part of the body from head to toe [1]. Hydatid cyst of the head and neck region, especially the skull base, is extremely rare. Cysts in this location can present with atypical clinical symptoms, as in our case, with lower cranial nerve paresis, such as palatal and vocal cord palsy,

Figure 2. Heavily T_2 weighted axial (a) and T_1 weighted sagittal (b) images show a large CSF signal multivesicular lesion (white arrows) with multiple septa representing the walls of the daughter cysts, giving a "bunch of grapes" appearance in the right posterior skull base. The lesion extends superiorly into the posterior fossa to lie in the inferior aspect of the right cerebellopontine cistern, causing mass effect on the medulla, right lower cranial nerves and ipsilateral vertebral artery.

Figure 3. Heavily T_2 weighted (a) axial and (b) coronal images show a large multilobulated cystic mass with daughter cysts showing thin low-signal-intensity peripheral rim ("rim sign"; white thin arrows) and irregular linear low-signal-intensity structures within the cyst, suggestive of separated collapsed membranes ("serpent sign") (white thick arrows).

mimicking a malignant mass [4]. Imaging has an important role in making a diagnosis of hydatid cyst. Hydatid cysts in unusual anatomical locations are difficult to diagnose on imaging, even in patients from endemic regions [1, 4, 8]. Therefore, familiarity with imaging findings is crucial to make a pre-operative diagnosis. On CT, this multiloculated cystic skull base lesion mimicked a malignant mass. MR is a more useful imaging modality when a complex cystic head and neck lesion is present [3, 9]. A cystic lesion with multivesiculated "bunch of grapes" appearance, a peripheral T_2 hypointense rim ("rim sign") and separated collapsed membranes as irregular low-signal-intensity linear floating structures ("serpent sign") were considered diagnostic for hydatid disease [1–3, 7]. These signs are best demonstrated on the heavily T_2 weighted sequence. Hydatid cyst should be considered in the differential diagnosis of cystic lesions in the head and neck region in endemic areas.

Conclusion

This report illustrates that hydatid cyst of the head and neck region may occasionally present with signs of lower cranial nerve compression. MRI should be considered as the diagnostic modality for hydatid disease. The heavily T_2 weighted sequence has been shown to be a very useful sequence in making a definitive diagnosis of hydatid cyst.

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