Interdural cavernous sinus dermoid cyst in a child: case report

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Interdural dermoid cysts (DCs) of the cavernous sinus (CS), located between the outer (dural) and inner layer (membranous) of the CS lateral wall, are rare lesions in children. The authors report on a 5-year-old boy with third cranial nerve palsy and exophthalmos who underwent gross-total removal of an interdural DC of the right CS via a frontotemporal approach. The patient had a good outcome and no recurrence at the 12-month follow-up. To the best of the authors' knowledge this is the second pediatric case of interdural DC described in the literature.

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KEY WORDS dermoid cyst; interdural cavernous sinus tumors; ophthalmoplegia; skull base surgery; pterional approach; oncology
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Case Report
History and Examination
A previously healthy 5-year-old boy presented with a 1-year history of bilateral frontal headache, chronic fatigue, diplopia, ptosis, and swelling of the right eye. Neuro-ophthalmological examination revealed best corrected visual acuity of 20/20 in both eyes, normal color vision, and right third cranial nerve paresis, together with a right exophthalmos, crossed diplopia, upper-eyelid ptosis, and anisocoria with right fixed mydriasis and absence of light pupillary reflex; results of the anterior segment and fundus examination were unremarkable (Fig. 1). A cranial CT scan disclosed a low-density, well-defined, smooth, oval lesion in the region of the right CS; there was a slight hyperdense rim into the cyst wall, perhaps due to calcifications and a scalloping of the sphenoid bone (Fig. 2). The MR images revealed a well-circumscribed oval lesion with smooth contours inside the lateral wall of the right CS. The lesion was hypointense on T1-weighted images, with some hyperintense signal at its periphery, and it was hyperintense and heterogeneous on T2-weighted images. There was no enhancement on T1-weighted images obtained after gadolinium injection, except for a slight rim around the edges of the lesion. The lesion displaced the intracavernous internal carotid artery (ICA) medially, and diffusion-weighted imaging (DWI) sequences revealed a restricted proton diffusion within the lesion (Fig. 3).

The differential diagnosis of the lesion included the following possibilities: DC of the lateral wall of the CS; arachnoid cyst; third, fourth, or fifth cranial nerve neurinoma; cystic glioma; or craniopharyngioma. The clinical and neuroradiological findings supported the first hypothesis—the diagnosis of DC of the lateral wall of the CS. Therefore, surgery was offered to the child’s parents as the best treatment because the chance of expected symptomatic improvement was estimated to be higher than the chance of surgery-related morbidity. The child underwent surgical removal of the lesion after his parents gave informed consent according to our institutional code of ethics.

Operation
After induction of general anesthesia and endotracheal intubation, venous and arterial accesses were established. The patient was placed supine and the head was secured in a Mayfield clamp and rotated 30° to the left and slightly extended, with the right shoulder lifted up. The head was coregistered to the Neuronavigation System (Treon Neuronavigation System; Medtronic, Inc.). A right frontotem-
poral craniotomy was performed after a curvilinear skin incision was made behind the hairline, separately cutting the temporal fascia and muscle to preserve the frontal branch of the facial nerve by retracting the fascia anteriorly and the muscle posteroinferiorly. The dura mater was opened with a standard C-shaped incision, and the arachnoid layer was microsurgically dissected to open the anterior half of the sylvian fissure by using a self-retractor. The right ICA and the right optic nerve were visualized, along with a whitish mass bulging through the lateral wall of the CS. The outer (meningeal) layer of the lateral wall was incised and opened in a stellate fashion over the most prominent part of the tumor, where no cranial nerves were identifiable at higher magnification and corresponding approximately to the Parkinson’s triangle area (Fig. 4).

The mass was located inside the lateral wall of the CS and then was progressively removed in a piecemeal fashion from the inner side of the capsule with no significant bleeding; it showed a typical appearance consistent with greasy material and hair tufts. At the end of gross-total removal the cranial nerves became visible through the inner meningeal layer of the CS lateral wall and through the inner DC capsule, which was preserved to avoid their injury. At the end of the excision, the surgical cavity was inspected with a microsurgical mirror to remove any remnant from inside. Hemostasis was achieved by packing the CS with oxidized cellulose, and the dura, the skull bone, and the skin wound were then closed in a standard fashion.

Postoperative Course and Follow-Up

The patient’s postoperative recovery was uneventful, and a CT scan performed 1 day after surgery excluded surgical complications. The patient was discharged 4 days after surgery with no new neurological complaints. Histopathological examination of the surgical specimen confirmed that the tumor was a DC with abundant keratinic material; numerous foreign-body giant cells and occasional hairs were also seen (Fig. 5). At the 2-month follow-up the third cranial nerve impairment had partially recovered: the child no longer complained of diplopia; proptosis and ptosis had disappeared; and there was still a right mydriasis with a weak light pupillary reflex and limitation in eye adduction. Six months after surgery, an MRI study (Fig. 6) confirmed gross-total removal with decompression of CS neurovascular structures and without any tumor remnant or relapse; the preoperative hyperintense signal in FLAIR images completely disappeared after surgery (Fig. 7). At the last follow-up visit, 12 months after surgery, the child showed an almost complete recovery of eye movements and light pupillary reflex, and the right mydriasis had disappeared.

Discussion

We report the second pediatric case of an interdural DC of the lateral wall of the right CS in a 5-year-old boy with a good outcome 1 year after gross-total removal. Dermoid cysts are extremely rare nonneoplastic tumors account-
Clinical findings in DCs of the CS usually occur quite late, due to their slow growth pattern and the fact that symptoms are caused by direct focal compression of adjacent neural structures or sometimes by the acute rupture of the DC capsule leading to aseptic chemical meningitis and raised intracranial pressure.5,10,12,16,21,23,29 The most frequent symptoms and signs may include headaches, oculomotor palsy, diplopia, visual loss and, more rarely, visual field defects, exophthalmos, hemiparesis, and raised intracranial pressure.2,7,16,20,22–24,29 Dermoid cysts located in the lateral wall of the CS typically cause different degrees of oculomotor palsy, especially of the third and fourth cranial nerves,2,16,19,20,22–24 along with dysfunction of the ophthalmic and maxillary division of the fifth cranial nerve, both as trigeminal neuralgia and hypesthesia or impairment of temporal and masseter muscles.2,24 The pupillary muscles are not spared because the parasympathetic fibers are very sensitive to compression, and sometimes are affected earlier than oculomotor muscles.18,20 Perrini et al.22 reported a case of an interdural DC of the CS with isolated sixth cranial nerve palsy, whereas Rato and colleagues23 described a patient reporting only headaches. Our patient suffered from bilateral frontal headaches, diplopia, right ptosis, and right exophthalmos. His medical history was unremarkable. At neurological examination he showed a typical isolated right third cranial nerve impairment leading both to right ptosis and exotropia, and to fixed miosis due to loss of light pupillary reflex (Fig. 1). There was right exophthalmos, probably due to CS venous congestion, albeit no sign of papilledema was present at fundus examination. There was no dysfunction of either the fourth or sixth cranial nerves or of the trigeminal nerve.

Neuroradiological investigations are crucial for diagnosis and for planning the right surgical strategy. A CT scan usually shows an interdural DC as a low-density, oval lesion with the same signal for fat or CSF without contrast enhancement,2,11,17,28 although hyperdense DCs with capsular calcification in their wall have been described.6 On MR scans DCs usually appear as hyperintense and hypointense lesions in T1- and T2-weighted images, respectively, without peripheral edema;2,7,16,17,23,28 on T2-weighted images the signal is frequently mixed and inhomogeneous, depending on the different proportions of DC elements.2,7,16,17,23,28 Use of FLAIR and DWI sequences may help in making the diagnosis of DCs10,16 by showing the cysts as hyperintense lesions and with typical restricted proton diffusion, respectively. The MR features suggesting CS interdural location of DCs include a smooth, oval, and regular shape and the medial displacement of the intracavernous ICA without its narrowing, as opposed to intracavernous and invasive CS tumors that usually encase the ICA.14,16,23,24 Differential diagnosis must consider trigeminal, oculomotor, and trochlear neurinomas; melanomas; metastatic tumors; cavernomas; arachnoid cysts; cystic gliomas; craniopharyngioma; and epidermoid cysts.7,14,23

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temporal plus zygomatic osteotomy, providing a wider exposure of the temporal lobe and of the CS surrounding the risks and benefits of pursuing a radical resection (Fig. 3).

The 2 most used surgical approaches for DCs of the CS are intradural frontotemporal (pterional), frontotemporal plus zygomatic osteotomy, and frontoorbitozygomatic (Table 1). The frontoorbitozygomatic route provides a wider exposure of the temporal lobe and of the CS region, reducing brain retraction, even though frontotemporal craniotomy can achieve enough working space and is less invasive, especially in children. On this basis we decided to perform a right frontotemporal craniotomy in our patient via an intradural approach to the CS region, which was easily exposed after a microsurgical opening of the anterior half of the sylvian fissure with minimal brain retraction by the self-retractor. The DC appeared as a whitish bulging of the CS lateral wall that was opened at the dome of the tumor, corresponding approximately to Parkinson’s triangle (Fig. 4), and where no cranial nerves were recognizable at higher magnification.

Regarding the extent of resection, there is a controversy surrounding the risks and benefits of pursuing a radical resection of interdural DCs together with their capsule. In their landmark series of 43 surgically treated patients with DCs and epidermoid cysts of the CS, Yaşargil et al. stated that radical removal of these lesions including the capsule increases the surgical morbidity significantly in terms of cranial nerve palsies, with no advantages for patients, given that the risk of DC relapse is quite low even after subtotal removal. Nevertheless, some authors have performed radical removal of DCs without reported morbidity. Given the high risk of cranial nerve damage in the case of strict adherence between the capsule and neurovascular structures of the CS, we preferred to perform a gross-total removal of the DC, aiming to obtain maximal debulking of the CS as described by other authors. After opening the outer (dural) layer of the CS lateral wall at the dome of the tumor (Fig. 4), we carefully removed all the contents of the DC in a piecemeal fashion until the cranial nerves appeared through the DC inner capsule. We then removed all the contents of the DC in a piecemeal fashion. The DC appeared as a whitish bulging of the CS lateral wall that was opened at the dome of the tumor, corresponding approximately to Parkinson’s triangle (Fig. 4), and where no cranial nerves were recognizable at higher magnification.

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Dermoid cysts have a thick capsule made of stratified squamous epithelium and contain dermal structures such as hair, hair follicles, sebaceous glands, teeth, or nails; they are usually filled with a yellowish-brown fluid as a result of sebaceous gland secretion and progressive epithelial desquamation. The cyst enlargement relies on the secretion and degeneration of dermal elements that progressively enlarge the capsule of the cyst, especially toward less resistant spaces like arachnoidal cisterns, sulci, or fissures, and its consistency depends on the ratio of

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<td>North et al., 1993</td>
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<td>Lat wall of CS, interdural</td>
<td>Isolated CN III palsy</td>
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<td>Nakagawa et al., 1997</td>
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<td>Akdemir et al., 2004</td>
<td>18/M</td>
<td>Lat wall of lt CS, interdural</td>
<td>HA, diplopia, ophthal- moplegia (CN III palsy), blurred vision</td>
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<td>Tun et al., 2008</td>
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<td>Lat wall of lt CS, interdural</td>
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<td>Partial recovery, improvement of diplopia &amp; ptosis</td>
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<td>Rato et al., 2012</td>
<td>21/F</td>
<td>Lat wall of lt CS, interdural</td>
<td>HA, blurred vision</td>
<td>Frontotemporal craniotomy</td>
<td>Total</td>
<td>6 mos, none</td>
<td>No complications</td>
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<td>Perrini et al., 2013</td>
<td>37/M</td>
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<td>No complications</td>
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<td>Present study</td>
<td>5/M</td>
<td>Lat wall of rt CS, interdural</td>
<td>HA, diplopia, CN III palsy, exophthalmos</td>
<td>Frontotemporal craniotomy</td>
<td>Gross-total</td>
<td>12 mos, none</td>
<td>No complications</td>
<td>Partial recovery, disappearance of diplopia &amp; exophthalmos, improvement of CN III palsy</td>
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CN = cranial nerve; FU = follow-up; HA = headache; NA = not available.
various elements within the wall. A DC differs from teratomas and epidermoid cysts in that teratomas contain tissue from all 3 germinal layers and not only from the ectoderm as in DCs, whereas epidermoid cysts are usually filled with waxy concentric lamellae of cholesterol crystals. Histopathological examination of the surgical specimen revealed the typical findings of DCs, such as abundant keratinic material and occasional hairs that originated from the ectodermal layer; numerous foreign-body giant cells were also present (Fig. 5).

Regarding surgery-related morbidity, after the seminal work by Yaşargil et al. who described transient cranial nerve palsies and aseptic chemical meningitis, more recent reports have not reported neurological deterioration in almost all patients. Our patient showed a significant improvement of third cranial nerve palsy 1 month after surgery and a reduction of exophthalmos. A postoperative MRI study performed 6 months after surgery confirmed gross-total removal of the DC and showed complete debulking of the CS lateral wall with its neurovascular structures (Fig. 6). The FLAIR images revealed a hypointense signal inside the surgical cavity as compared with the previous hyperintense lesion (Fig. 7); there was no sign of tumor remnant or relapse. At the last follow-up visit 12 months after surgery, diplopia and exophthalmos had completely disappeared, whereas there was still a slight third cranial nerve impairment at neurological examination.

Conclusions

Interdural DCs of the CS are rare intracranial lesions, especially in pediatric patients, but they should be considered in the differential diagnosis of CS tumors. Clinical findings consist of typical CS syndrome with third, fourth, fifth, and sixth cranial nerve impairments of various degrees. Neuroradiological workup enables us to recognize interdural DCs of the CS due to their typical appearance of smooth, oval lesions without contrast enhancement inside the lateral wall of the CS on MRI and with typical restricted proton diffusion on DWI sequences. The aim of surgery is to achieve gross-total removal while preserving the neurovascular structures, although some authors suggest a more aggressive resection including the DC capsule. In this report we have described a 5-year-old boy with an interdural DC who underwent gross-total removal through a frontotemporal craniotomy, who had a favorable outcome and no tumor recurrence at the 1-year follow-up. To the best of our knowledge this is the second pediatric case of interdural DC of the CS reported in the literature.

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Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Giordano. Acquisition of data: Giordano, Peri, Bacci, Basile, Guerra, Bergonzini, Buccoliero, Spacca, Iughetti. Analysis and interpretation of data: Giordano, Spacca. Drafting the article: Giordano, Spacca. Donati. Critically revising the article: Giordano, Spacca. Reviewed submitted version of manuscript: Giordano. Approved the final version of the manuscript on behalf of all authors: Giordano. Study supervision: Giordano, Genitori.

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