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CASE REPORT

# Intramedullary Epidermoid Cyst of the Conus Medullaris: A Case Report and Literature Review

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**Background:** Central nervous system tumors are usually located in the brain, and spinal cord tumors account for approximately 20% of central nervous system tumors. Epidermoid cysts constitute <1% of all intraspinal tumors. It consists of squamous epithelial-lined cysts containing keratin, cholesterol, and cellular granules. Epidermoid cysts can be classified as congenital, acquired, extradural, extramedullary, or intramedullary according to etiology and location. The intradural intramedullary type is uncommon.

**Case Presentation:** An 11-year-old female patient had back pain with radiation to both lower extremities but worsened on the left side for 5 years. On neurological examination, motor strength was 3/5 for hip flexion and knee extension bilaterally and 5/5 for other key muscle groups. Contrast-enhanced lumbosacral magnetic resonance imaging (MRI) revealed T1 hypointense and T2 hyperintense lesions in the L2-L4 intramedullary conus and cauda equina. Laminectomy and near total resection were done, and histopathological examination revealed an epidermoid cyst. On the third postoperative day, the patient was discharged with completely resolved back pain and an improvement in lower extremity motor power. At monthly follow-up visits for a further 6 months, her back pain and weakness completely resolved, and she had no neurologic deficits. A postoperative lumbosacral MRI was done and confirmed near total excision of the tumor.

**Conclusion:** Intramedullary conus epidermoid cysts are rare but not unknown to neurosurgeons. MRI with diffusion-weighted images (DWI) is an imaging modality of choice. Asymptomatic patients can be conservatively treated. Once the patient has progressive symptoms and signs of compression, surgical excision is recommended. Meticulous electrocauterization may help decrease tumor regrowth in the remnant capsule, which is recommended. Avoiding leakage of cyst fluid into the subarachnoid space helps to avoid postoperative chemical meningitis. Radiotherapy is an option for the management of multiple recurrences.

**Keywords:** epidermoid cyst, conus medullaris, cauda equina, intramedullary

## Introduction

Epidermoid cysts were first described by Couvelaire in 1835. It means "perlee tumor" (pearly tumor). Epidermoid cysts are benign, slow-growing lesions that account for <1% of all intraspinal tumors. It can arise congenitally in patients with spinal dysraphism, such as spina bifida, dermal sinus, and syringomyelia, whereas acquired causes include recurrent lumbar punctures resulting in the implantation of ectodermal cells in the spinal canal. Epidermoid cysts are rarely identified in the cervical and upper thoracic regions compared to the conus, with a slight female predominance. Extramedullary epidermoid cysts outnumber complete intramedullary lesions, which is uncommon in clinical practice (Alain Roux, March 1992).

## Case Presentation

An eleven-year-old female patient with lower back pain for five years associated with bilateral radiation to the lower extremities, which is more pronounced on the left side. The pain was insidious in onset, worsening over a 2-month period. On the neurological exam, motor power became 3/5 on hip flexion and knee extension; the other lower extremity muscle power was 5/5; the tone was normotonic; and reflexes had been +2 in all deep tendons. She was referred to our

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center for surgical intervention; otherwise, she had no history of spinal trauma or any spinal procedure, including a lumbar tap.

Laboratory examinations on the day of admission included a complete blood count and biochemical studies, which showed no apparent abnormalities. Contrast-enhanced lumbosacral magnetic resonance imaging (MRI) showed type 1 (T1) hypointense and T2 hyperintense, 6 cm cranio-caudally and 2 cm antero-posteriorly, expansile lesions that extended from L2–L4 with involvement of the conus and cauda equina (see Figure 1). It shows marginal enhancement after contrast administration. In the operating room, under general anesthesia, a prone position was made and a vertical skin incision was made to expose the operative area. An L2–L4 complete laminectomy with L1/L5 partial laminectomy is done as the tumor extends in both the cranial and caudal poles and the dura opened on the midline. Intraoperatively, the mass anchored the conus intradurally, filling the spinal canal and displacing the caudal roots anteriorly and laterally, with adherence to the nerve roots on each aspect from L2–L4 (Figure 2C–D). Surgical microscopic examination revealed a subpial thin-walled cyst with a grayish content. We did no longer have intraoperative motor evoked potential (MEP), sensory evoked potential (SEP), electromyography (EMG), or cavitron ultrasonic surgical aspirator (CUSA) for debulking; for this reason, we did no longer use them in this case either. Tumor dissection started at the posterior median sulcus. Few reviews have advocated the use of ultrasonography before myelotomy in difficult instances. The whitish cyst contents are expelled effortlessly because they were suckable; the cyst wall was excised in part, and the capsule, which was firmly adherent to the spinal cord parenchyma, was left in a few areas to not sacrifice neural tissue in an attempt to reap total capsulectomy. Care taken to prevent spillage of the cyst into the subarachnoid area. A postoperative lumbosacral MRI confirmed near total excision (see Figure 3). The jelly-like cyst content and its wall were sent for histopathological examination, which confirmed a fragmented fibrous cyst wall coated through maturing stratified squamous cells with scattered keratin flecks and fragments of bland skeletal muscle and adipose tissue, which are typical of epidermoid cysts (Figure 4). The postoperative path was uneventful; the pain had resolved, motor and sensory function were intact, and the patient was discharged on the third postoperative day. She had been being followed in our outpatient clinic for the last 6 months; throughout the follow-up, the pain had completely resolved, the neurologic exam became uneventful.

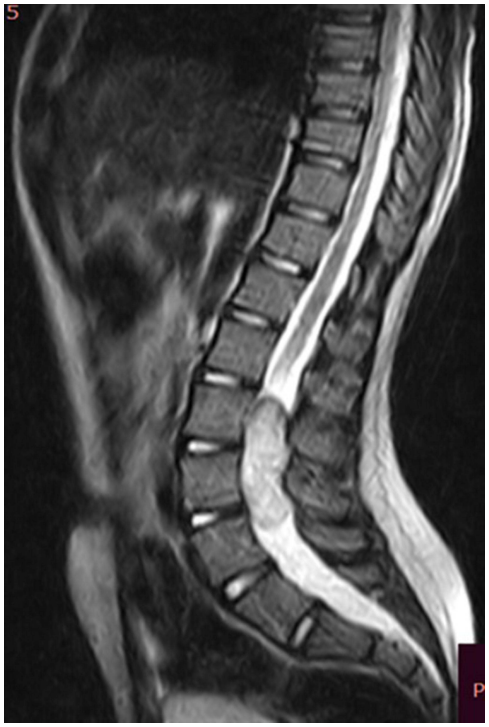


**Figure 1** Lumbosacral MRI of the patient preoperatively done sagittal T2 image demonstrating hyperintense lesion extending from L2-L4.

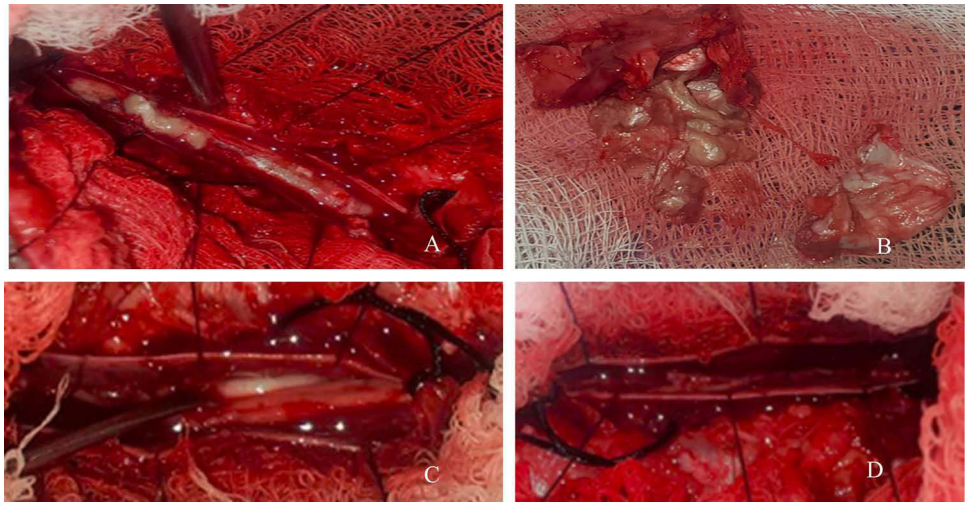


**Figure 2** (**A**–**D**) Intraoperative picture demonstrating tumor after dural opening and myelotomy, tumor samples and complete tumor removal.

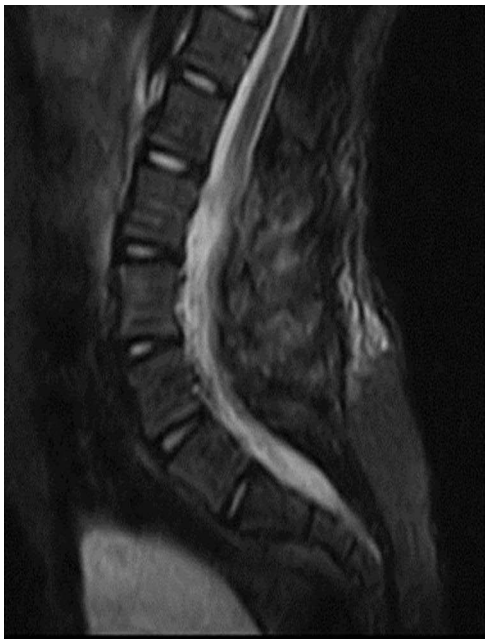

**Figure 3** Postoperative control lumbosacral MRI demonstrating complete tumor excision.

## Discussion

The pathogenesis of epidermoid cysts of the spine can be divided into two types: congenital and acquired. They are also classified according to segmental distribution, associated congenital lesions, and clinical presentation. Acquired epidermoid lesions are discovered years after one or more lumbar punctures and are thought to be the result of iatrogenic skin penetration. Van Gilder and Schwartz describe an animal model that produces epidermoid cysts and dermoid cysts in mice by direct transplantation of homogeneous skin along the neuroaxis. It is thought that congenital epidermoid cysts usually arise from ectodermal inclusions that change in early fetal life and may be associated with the closure of the neural tube. There are few or no reports of intramedullary epidermoid cysts in the cervical region of the spinal cord. The most common location of these cysts is the thoracic region, especially between T5 and T8[1] (see Table 1). There is an association between intramedullary epidermoid cysts and bone and skin defects.[1] However, it is less common than dermoid or extramedullary epidermoid cysts. Dermal sinuses can also cause meningitis in children, resulting in the early

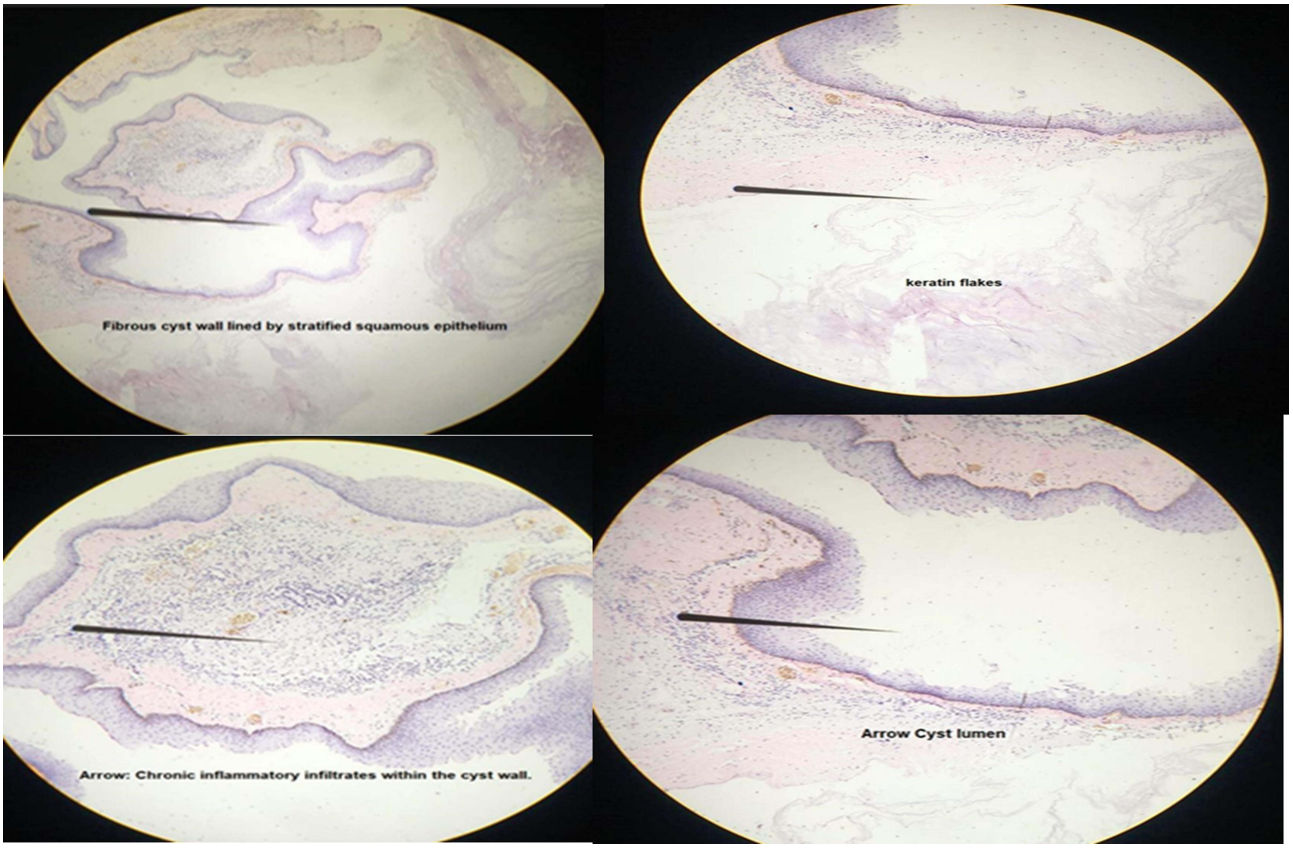


**Figure 4** Histopathology microscopic pictures demonstrating keratin flakes, fibrous cyst wall lined by stratified squamous epithelium, chronic inflammatory infiltrates within the cyst wall and a cyst lumen.

detection of intramedullary epidermoid cysts. The association between epidermoid cysts and different spinal cord disorders, including diastematomyelia or syringomyelia, has not been reported frequently.[1] Our patient had no evidence of spinal cord congenital disorders, including scoliosis, tufts of hair, skin dimpling, or skin pigmentation.

Intramedullary epidermoid cysts were found in patients with a history of lumbar puncture, while our patient had no spinal surgery record.[1] Lumbar epidermoid cysts were reported in one patient with a history of more than 70 spinal cords and in another 24-month-old patient with a history of lumbar puncture.[5] In addition, one patient was diagnosed with an epidermoid cyst after disc herniation surgery.[5] However, approximately 40% of these cases are idiopathic in nature.[5]

Epidermoid cysts account for less than 1% of all intraspinal tumors.[1] Intracranial epidermoid cysts, although rare, are more common with a skull-to-spine ratio of 1:14. Spinal epidermoid cysts are usually intradural extramedullary cysts, whereas only 61 true intramedullary epidermoid cyst cases have been described in the literature to date.[2] Epidermoid cysts in the spinal cord can be classified as epidural, intradural, extramedullary, or intramedullary cysts. Of these, intradural and intramedullary variations are rare. This is the fifth report of an intramedullary epidermoid cyst in a child.

Epidermoid cysts are lined with stratified squamous epithelium and supported by a layer of collagen. Keratin is released slowly and is cleaved by epithelial cells in the direction of the cyst, resulting in smooth white tissue rich in LDL cholesterol crystals forming concentric lamellae, often described as waxy. Compared to the multi-tone growth rate found in most tumors, growth is close to that of normal skin. Malignant transformation to form local tumors has been documented but is rare. Biochemical analysis of cyst material shows too much fat, but cholesterol levels are not high. Loose fatty acids and cholesterol can be released by inflammatory cells in response to the necrosis of degenerated squamous cells. The rupture of epidermoid cyst contents into the subarachnoid space may also cause granulomatous meningitis or chemical meningitis due to the release of fatty acids and/or low-density lipoprotein[1,5] (see Figures 3 and 4).

Except for tumors associated with a dermal sinus, intramedullary epidermoid cysts are very slow-growing tumors that usually occur after the second decade of life.[1] Patients aged 3 to 71 years, median age 34.[1] The onset and duration of symptoms ranged from 2 days to 53 years (mean 6 years) (see Table 1). In this decade, with the spread of modern diagnostic tools, such as tomography and magnetic resonance imaging, the time between symptoms and diagnosis has shortened. However, this guarantee may not apply to our site, as such images are not always easy to access. Therefore, the patient had low back pain and weakness in the lower extremities until five years before the diagnosis. Signs and symptoms are similar to those of most spinal cord diseases[1] (see Table 1).

The clinical presentation of epidermoid cysts of the spine depends on the size of the tumor and its location in the spine. Back and leg pain are the most common signs and symptoms, along with focal neurological deficits. Other symptoms include weakness, muscle cramps, numbness, paresthesia, and bowel and bladder problems.

Cysts can rupture, causing signs and symptoms to appear suddenly. Cyst rupture may be due to increased pressure due to Valsalva maneuvers, such as coughing and sneezing.[5] Because of the slow progression of the disease, which is the main cause of delayed presentation, patients usually present in early or middle adulthood.[7]

Plain radiographs of the spine may also show evidence of chronic intraspinal lesions. Myelography is useful in assessing the mass effect on the spinal cord, intradural filling disorder, or subarachnoid obstruction.[8] It usually checks for the size and length of the tumor but does not reveal its nature.

CT scans show the shape, length, and density of epidermoid lesions and can generally give prognostic evidence. Epidermoid cysts are characterized by homogeneous hypodense lesions with well-defined borders; they usually do not enhance. Calcifications can be seen in these tumors. An MRI can also provide an accurate diagnosis. Epidermoid cysts often occur with variable signals on the MRI and sometimes different signals within the same cyst. Other features include the absence of edema in the surrounding tissue, clear borders, and calcifications.

After an intravenous injection of gadolinium, sometimes edema can be seen peripherally. The difference in signal intensity may reflect the difference in lipid and protein content between these organisms. T1 and T2 weighted signals on MRI ranged from hypointensity to isointensity. In some cases, hyperintensities on T2-weighted images were described.

**Table 1** Previous Cases with an Intramedullary Epidermoid Cyst

| No. | Authors | Age/Sex | Presentation and Duration of Symptom | Tumor Location on the Spine | Tumor Size | Management | Outcome |
|---|---|---|---|---|---|---|---|
| 1 | (Christina Fleming, 2011)[2] | 24/f | Recurrent back pain and right foot weakness of 5 years | Conus medullaris | 5 cm | L1/L2 laminectomy, exploration and debulking | Bilateral lower extremity weakness and persistent back pain |
| 2 | (Rafael Cincu, 2007)[3] | Case 1; 27/m | Lower extremity weakness of 1year | ThoracicT5-T6 intramedullary | 1.8 cm | T4-T7 laminectomy and near total excision | Lower extremity weakness remained the same |
| | | Case 2; 28/f | Lower extremity weakness and urinary incontinence | Conus-epiconus region | Not specified | T12-L2 laminectomy resection | Improved |
| 3 | (Siddartha Reddy Musali, 2019)[4] | 6/f | Lower extremity weakness 5 months | Thoracic T9-T12 intramedullary | 4.9 cm | T9-T12 laminectomy, excision not specified | Improved |
| 4 | (ALAIN ROUX, march, 1992)[1] | 6/f | Lower extremity weakness and nocturnal enuresis/6 months | Thoracic T5-T6 intramedullary | Not specified | Laminectomy and near total excision | Improved |
| 5 | (Aseri, 2023)[5] | 17/m | Back pain 2months | Lumbar L5 intradural extramedullary | Not specified | L5 laminectomy and excision | Improved |
| 6 | (Kulwant Singh, 2016)[6] | 20/f | Lower extremity weakness/6 months | Thoracic T7-T9 intradural extramedullary | 3 cm | Gross total resection but laminectomy not specified | Improved |

Diffusion restriction on DWI is not uncommon in epidermoid cysts and helps differentiate epidermoid cysts from arachnoid and dermoid cysts.[1,7]

Asymptomatic epidermoid cysts can be treated conservatively; however, for those that show compression of the neural elements on their radiographs and have neurologic signs and symptoms, surgical intervention is recommended. In the report of a 63-year-old Japanese girl, internal decompression with cyst content aspiration was chosen as the surgical treatment because the cyst wall is tightly adhered to the roots, and they advocate this method in elderly patients and in congenital conditions due to slow regrowth.[4] Complete removal of the cyst is sure to prevent a recurrence. Sometimes, if the capsule is firmly attached to the nerve, trying to remove the cyst wall completely may cause damage to the nerve.

Therefore, en bloc resection is not possible in most cases. In such cases, cyst drainage and epithelial wall electrocoagulation are promoted to delay recurrence. Wall coagulation should be done carefully to avoid creating new neurologic deficits and worsening the already existing ones after surgery.[9,10] Intraoperative electrophysiological monitoring, including SEP, MEP, and EMG, is an important tool for microneurosurgery to intervene in a variety of intramedullary diseases. Intraoperative root stimulation may be helpful before myelotomy.

The coagulation of pial capillaries should be reduced.[11] Short-term administration of steroids (for 3 days) may also help reduce the signs and symptoms of chemical meningitis if cyst fluid enters the subarachnoid space.[11] Most authors credit Matson (cited from Rand and Rand2S) for emphasizing the importance of not attempting to complete the capsule resection when the cyst wall is tightly adhered to the neural tissue. In this case, attempts to remove the entire cyst wall are ineffective and can lead to rapid neurological complications.

Affected individuals are known to be at risk of recurrence if only one cell remains. This risk exists for a period equal to the age of the patient at diagnosis plus nine months.[5] Relapses have been reported; however, even partial excision of the intramedullary epidermoid cyst may improve the signs and symptoms in the long term and enable the patient to return to work. If these symptoms occur, another surgery is an option.[1] In 1977, a 17-year-old woman had to undergo three surgeries to remove this type of cyst. Ommaya reservoirs were placed inside the cyst for future flow. However, the risk of infection with this application was as high as 6%. There were also reports of seven-time recurrences of epidermoid cysts in the conus area, and the affected patient was still experiencing bilateral weakness and requiring pain management. Surgical treatment of epidermoid spinal cysts is difficult due to the difficulty of complete removal of the cyst wall and therefore the high risk of recurrence.[2] The prognosis is good, with recurrence depending on excision.[7]

In our case, the result took 6 months, which is a short-term result. The pain and weakness of the lower limbs improved completely. In previous studies, the recurrence rate of epidermoid lesions was approximately 10% to 29%.[8] In 1968, Stevens and Schlesinger described the first patient with a spinal epidermoid cyst and began radiation therapy. A 47-year-old male patient was seen with complaints of bowel and bladder incontinence for 11 years and received three courses of radiotherapy for suspected astrocytoma. Complete surgery was performed for the fourth time, and histopathological examination revealed an epidermoid cyst. Recently, Bretz et al reported a case of recurrent epidermoid cyst treated with surgery repeatedly. After radiation therapy, researchers describe a 6-year follow-up with no evidence of recurrence, and they noticed clinical improvement after the radiotherapy.[10]

## Conclusion

Intramedullary conus epidermoid cysts are rare but not unknown to neurosurgeons. MRI with DWI is an imaging modality that helps differentiate it from other lesions, such as arachnoid cysts. Asymptomatic patients can be conservatively treated. Once the patient has progressive symptoms and compression signs on imaging, surgical excision is recommended, but struggling to remove the last and most adherent capsule will worsen postoperative neurologic deficits. In these cases, electrocauterization helps prevent regrowth. Making every precaution to avoid leakage of cyst fluid into the subarachnoid space helps to avoid postoperative chemical meningitis, and if it occurs, short, course steroids may help in relieving the symptoms. For multiple recurrences, radiotherapy is an option.

## Abbreviations

CBC, complete blood count; CSF, cerebrospinal fluid; CT, computerized tomography; MRI, magnetic resonance imaging; T1W, T1-weighted images; T2W, T2-weighted images; DWI, diffusion-weighted images; L1, first lumbar

vertebra; L2, second lumbar vertebra; L4, fourth lumbar vertebra; L5, fifth lumbar vertebra; CNS, central nervous system; EMG, electromyography; MEP, motor-evoked potential; SEP, sensory-evoked potential; CUSA, cavitron ultrasonic surgical aspirator.

## Informed Consent

The participants' parents consented to the publication of this case detail and accompanying images.

## Disclosure

The authors declare that they have no conflicts of interest.

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