Pathology in Family Practice

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A 10-MONTH-OLD boy presented because his parents had noted a mass in the lower back that increased in size over the last 4 to 5 months. On physical examination, a nevus flammeus was noted over the lumbar region of the back, with a 1×1-cm firm mass beneath it in the midline. A small dimple in the skin was seen at the center of the mass (**Figure 1**). History and results of the remainder of the physical examination were not contributory.

Magnetic resonance imaging and computed tomographic scans showed a cystic lesion at L-4 connected to the skin by a sinus tract. The mass was both intradural and extradural (**Figure 2**).

The patient's family agreed to elective surgical removal of the tumor; the patient did well postoperatively and was discharged home.

Microscopic sections are shown in **Figure 3** and **Figure 4**.

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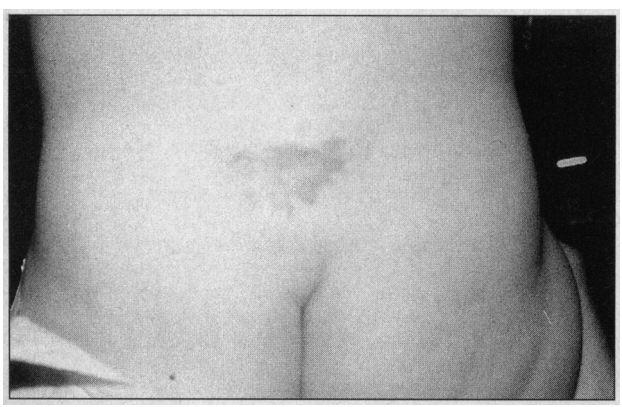

Figure 1.

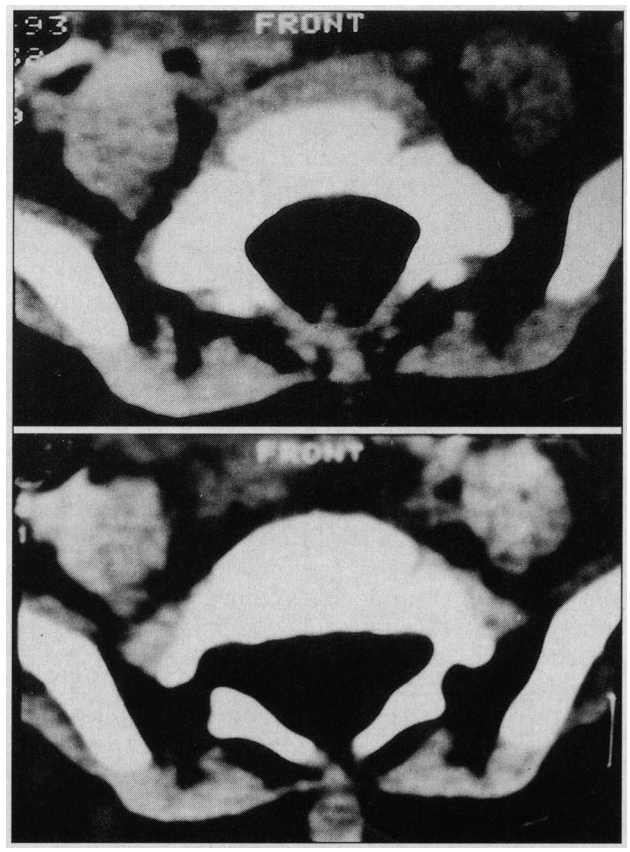

Figure 2.

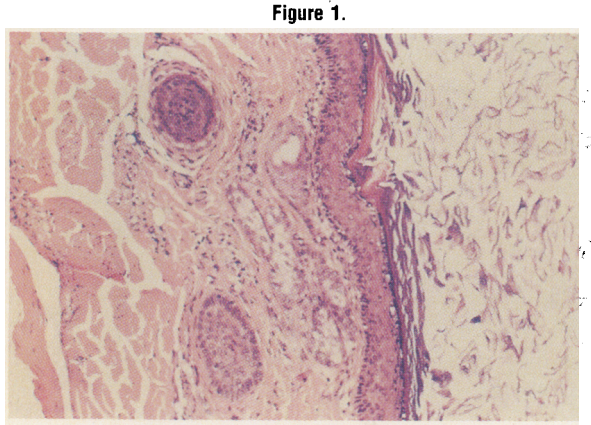

Figure 3.

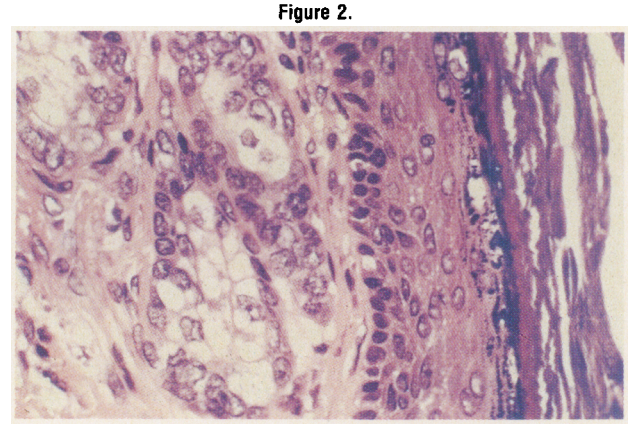

Figure 4.

Diagnosis and Discussion

Intraspinal Dermoid Cyst

Figure 1. *Photograph of patient showing nevus flammeus and mass with dimple.*

Figure 2. *Top, Computed tomography at level L-4 showing sinus tract and intraspinal cyst. Bottom, Computed tomography cut one vertebra higher showing extension of cyst cephalad.*

Figure 3. *Cyst containing keratin debris and lined by squamous epithelium, with underlying dermis containing sebaceous glands and hair follicle (hematoxylin-eosin, original magnification ×100).*

Figure 4. *High-power view of section seen in Figure 3 showing the squamous epithelium lining and the sebaceous glands (hematoxylin-eosin, original magnification ×400).*

Intraspinal dermoid cysts are slow-growing congenital tumors originating from inclusion of ectodermal cells during closure of the neural tube at about 3 to 5 weeks' gestation.[1] They are most frequently identified in the first two decades of life, the most common site of involvement being the lumbosacral region.[2] Other sites within the central nervous system include the inferior surface of the frontal lobes, the cerebellar vermis, and the fourth ventricle. Dermoid cysts make up 1% to 10% of intraspinal tumors.[3,4]

Spina bifida occulta is a commonly associated abnormality. An important and sometimes diagnostic feature is the finding of a dermal sinus tract leading from the skin, which may serve as a route for pyogenic infections.[2,5] This condition is a leading cause of recurrent meningitis in children. Another feature is the release of the contents of the cyst into the cerebrospinal pathway, which may lead to a rapidly fatal granulomatous form of meningitis. Although tumors may be either extradural or intradural, over 75% of the cases are extradural.

Macroscopic examination of dermoid cysts typically reveals a well-defined round or oval mass that varies greatly in size and wall thickness. The cyst characteristically contains sebaceous and keratin debris in which variable numbers of hairs may be entangled. Microscopic examination shows a cyst lined by simple squamous epithelium and supported by collagen-containing skin appendages such as sebaceous glands and hair follicles.[6]

A wide range of cysts may be encountered within the central nervous system, including arachnoid, neuroenteric, epidermoid, and teratomatous cysts.[7,8] Arachnoid cysts are frequently located in the sylvian fissure and are lined by collagenous connective tissue. The neuroenteric cysts are predominantly seen in the cervical region and are lined with columnar epithelium. Epidermoid tumors usually occur later in life, most commonly intracranially, and are lined with squamous epithelium alone. If the tumor contains elements from all three germ layers, then it is a teratoma.

Following surgical intervention for removal of the tumor, the prognosis is very good. Even when the capsule of the cyst must be left in place, which in some cases may be inevitable owing to tight adherence to the cord or cauda equina, the prognosis is good, with reoperation recommended only if recurrent growth of the tumor causes new symptoms.[1]

Selected from Arch Pediatr Adolesc Med. *1995;149:701-702. Pathological Case of the Month.*

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