A CASE OF CERVICAL SPINAL TERATOMA ASSOCIATED WITH DERMAL SINUS TRACT: MRI AND SURGICAL FINDINGS

Mehmet E. Üstün, M.D., Ahmet Erdoğan, M.D., Mustafa C. Avunduk, M.D.

Ankara University, Faculty of Medicine, Department of Neurosurgery (MEÜ, AE) and Pathology (MCA), Ankara - TÜRKİYE

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SUMMARY:

Intradural spinal teratomas are rare. We report a case in a 23-year-old man in whom Magnetic Resonance Imaging (MRI) revealed an intradural extramedullary tumour between the fourth and fifth cervical levels with a dermal sinus tractus. Histological examination showed a variety of tissues, including elements of all three germinal layers.

KEY WORDS:

Cervical vertebrae, Magnetic Resonance Imaging, Myelography, Spinal Cord Neoplasms, Teratoma

INTRODUCTION:

Cervical localization of a benign intradural extramedullary spinal teratoma with a congenital dermal sinus is rare. In a review of the literature we have not found any Magnetic Resonance Scan (MR-scan) of an intradural extramedullary spinal teratoma at this level with a congenital dermal sinus. We present here the myelography and MRI findings of our case with operative photographs and histological sections.

CASE REPORT:

A 23-year-old man was admitted with complaints of neck pain with muscle weakness that spread from the lower to the upper extremities in one week and progressive inability to walk. On examination there was a dermal sinus ostium in the skin overlaying the spine of the fifth cervical vertebra. He developed urinary hesitancy without incontinence. On neurogical examination there was a spastic quadriparesis, Hoffmann signs and hypoaesthesia below the cervical 4 level bilaterally, also impaired proprioception and vibratory sensation in the upper and lower extremities bilaterally. Superficial abdominal and cremasteric reflexes were absent.

Cervical spinal x-rays showed bifid spinous process at the fifth and sixth cervical levels.

Myelography performed by the lumbar route disclosed a total block between the C5-6 level (Fig. 1).

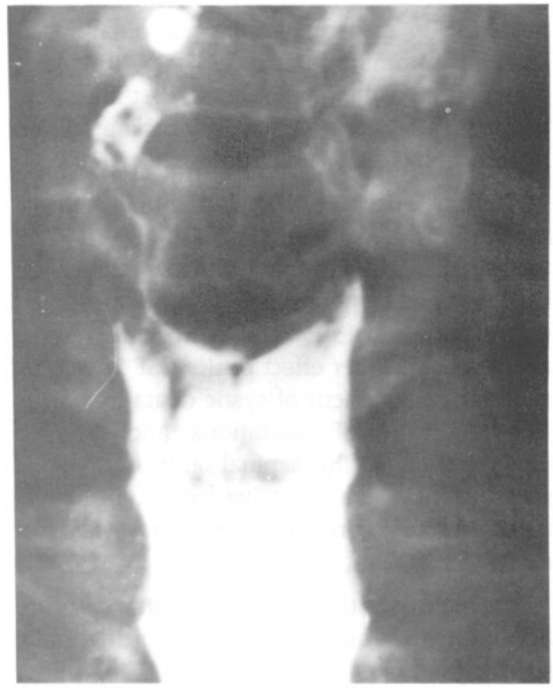

Fig 1 : Myelography showing a total block between the cervical 5- 6 level.

MRI showed an intradural extramedullary mass which had a dimension of approximately 2.5x1.5x2 cm between the fourth and fifth cervical levels. The lesion appeared hyperintense on T2-weighted images (Fig. 2) and hypointense on T1-weighted images with a dermal sinus tractus (Fig. 3).

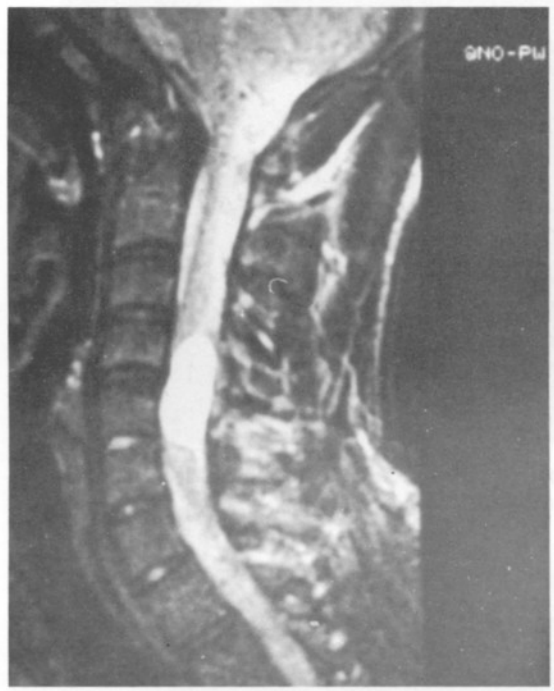

Fig 2 : T2-weighted image of the intradural extramedullary cervical mass.

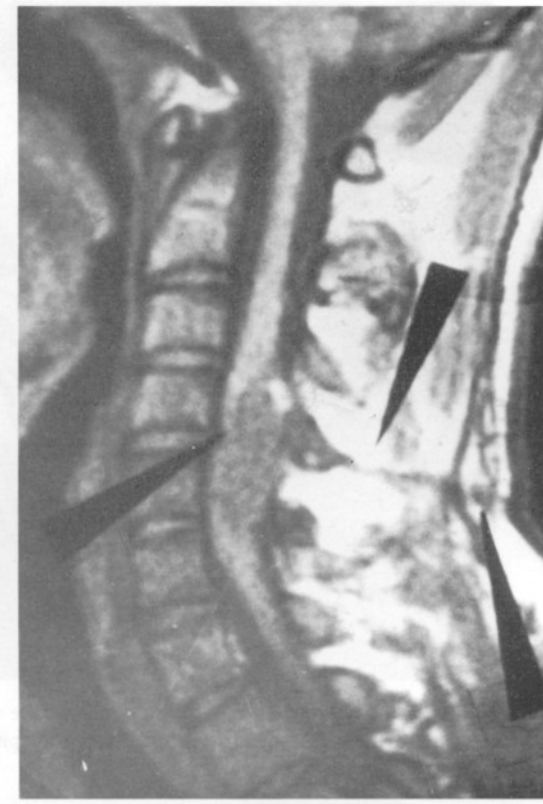

Fig 3 : T1-weighted image of the tumour with a dermal sinus tractus indicated by arrows.

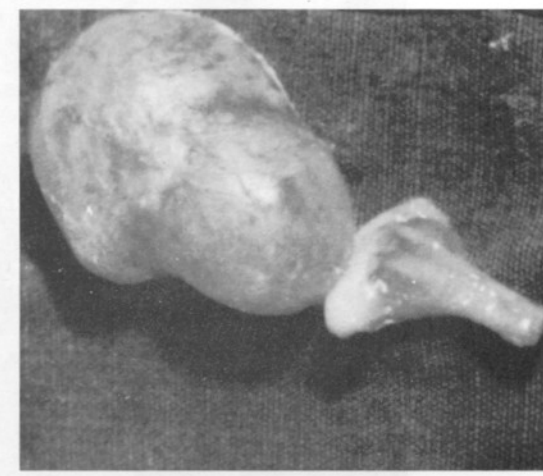

Fig 4 : Teratoma with the intraspinal portion of the dermal sinus tractus.

On exploration of the spine via C4, C5 total, and C3, C6 partial laminectomy, spina bifida deformity was seen at C5, C6 and a dermal sinus tractus that ascended from C5 to C4 and entered the spinal canal through C4 lamina was observed. After laminectomy it was seen that the sinus penetrated the dura and terminated in a cystic extramedullary tumour. The tumour, measuring approximately 3x2x1.5 cm, was totally removed and the dermal sinus tractus was resected (Fig.4).

Histological examination of the cyst wall showed it to be lined by epithelium varying from pseudostratified ciliated columnar to simple cuboidal (Fig.5). A layer of fat and fibrous connective tissue underlying the epithelium contained tubular glands possibly belonging to the intestinal canal (Fig.6) and also included smooth muscle bundles and rhabdomyoblasts (Fig.7). Under this layer mature cartilage was seen (Fig.6). No immature or malignant cells were present. As the tumour included elements of all three germinal layers and did not show cytological evidence of malignancy it was termed benign teratoma.

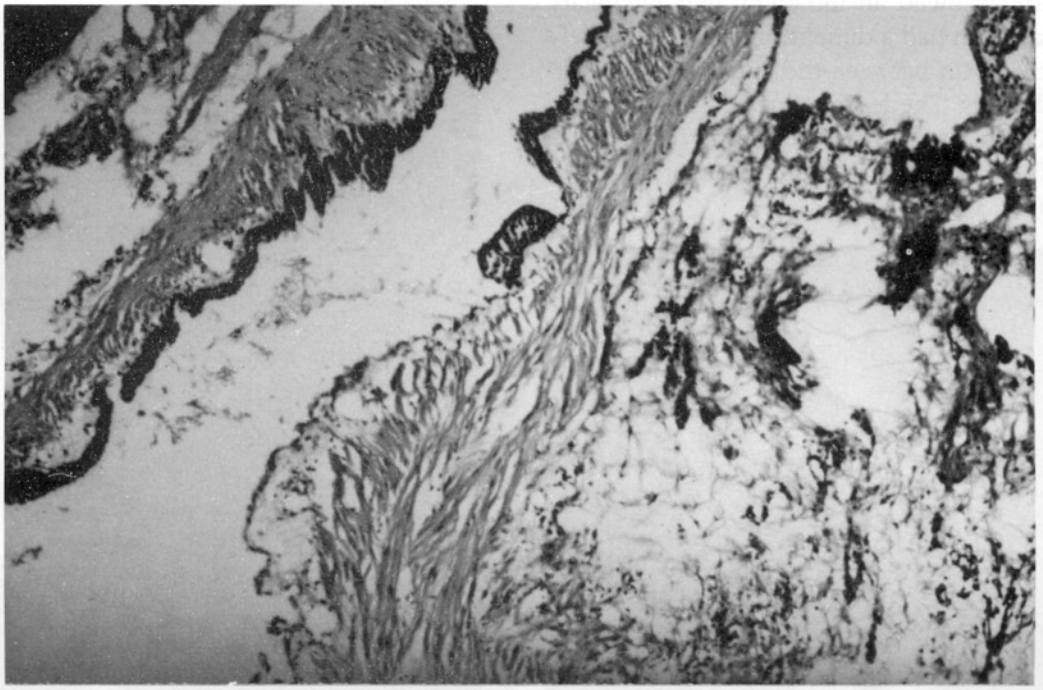

Fig 5 : Large cavity covered by transitional and simple cuboidal epithelium. A layer of smooth muscle bundle and fibrovascular tissue is seen underlying the epithelium (Hematoxylin Eosin (HE): x4).

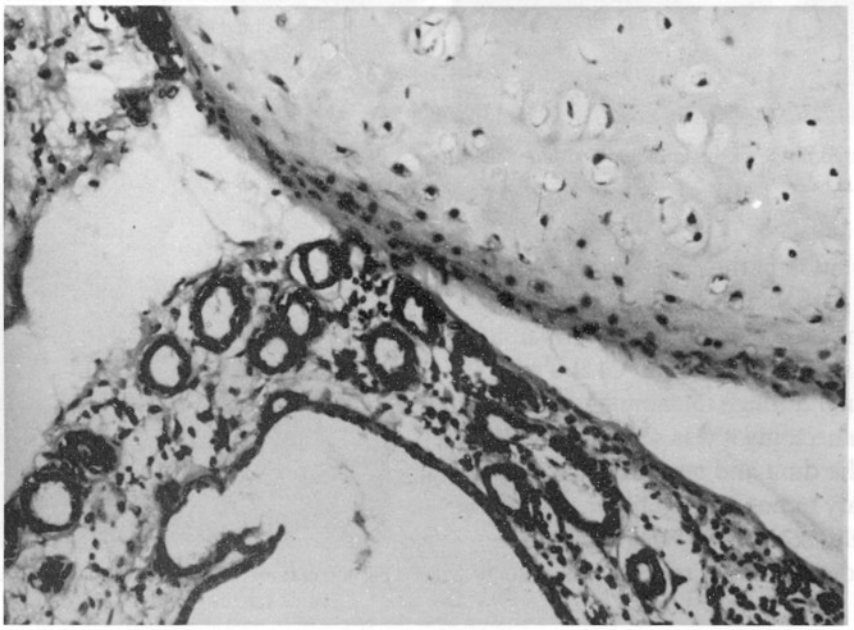

Fig 6 : Near the cartilage tissue there are tubular glands that may belong to the intestinal canal. A cyst wall covered by stratified epithelium is seen (HE: x10).

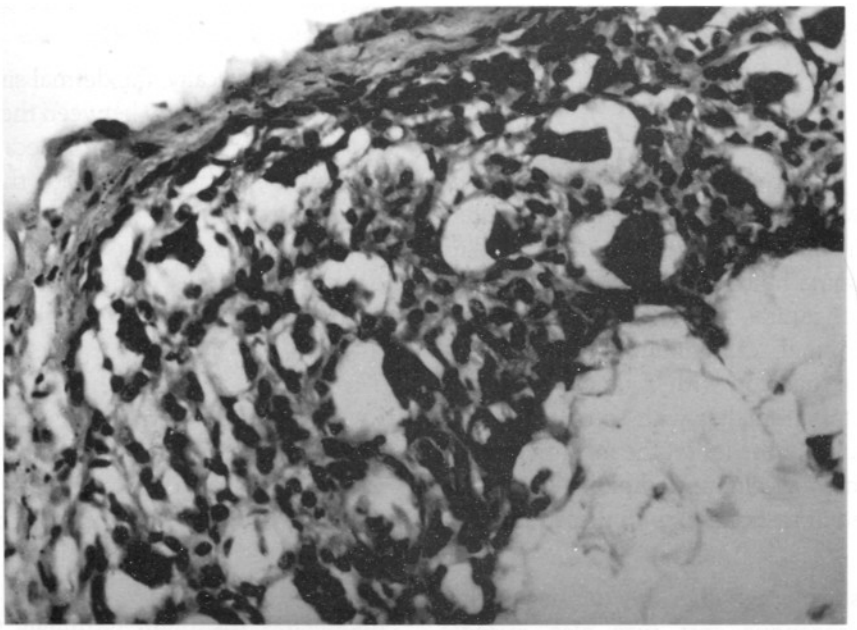

Fig 7 : Rhabdomyoblasts are seen in large lacuna (HE: x10).

On postoperative neurological examination there was an improvement in the quadriparesis and hypoaesthesia. At the time of discharge, one week after surgery, the patient had full strength in the upper extremities and was walking with help. The proprioseptive loss did not return. He was able to void spontaneously.

DISCUSSION:

Teratoma of the spinal cord is very rare, 53 cases have been reported, the first by Gowers (1876), and Monajati (1986) reported a benign intradural teratoma of the lumbar spine with MRI (4) and Hamabuchi (1989) a thoracolumbar intraspinal teratoma with CT scans (2).

Teratomas usually occur in the midline but may be found laterally and in various sites. The origin and nature of teratomas of the spinal cord are not yet clear, nor is the classification of these lesions uniform (5). Sachs and Horrax (6) noted that, in many of these tumours, derivatives of one or two germinal layers tend to overgrow the others and the total number of germinal layers may be difficult to ascertain. We agree with these authors that tumours with recognisable tissue from only two germinal layers should be termed "teratoids", and that only those with identifiable tissue from all three germinal layers should be labelled "teratoma". Willis (8) defines "teratoma" as "a true tumour or neoplasm composed of multiple tissues of kinds foreign to the part in which it arises". The most common components are skin, teeth, neural tissue, respiratory and alimentary mucosa, and glands. Although many authors have referenced this definition in their own discussions, it does not require the presence of all three germinal layers to make the diagnosis of teratoma. Hence, a number of bigerminal tumours (teratoids) have been included in the literature as teratomas (6). Furtado and Marques, in 1951, claimed that classification of germinal tumours as mono-, bi-, or trigerminal merely represented "a confession of inadequate examination". They believed that, if multiple serial sections covering the entire tumour were obtained, elements originating from all three germinal layers would be identified.

We reviewed all reported cases of intradural tumours termed teratomas, teratomatous cysts, cystic teratoid tumours, teratoid cysts, cystic teratomas, and teratoid tumours. From the histological descriptions, we could identify only 23 lesions that were trigerminal and could properly be termed intradural "teratoma". With the

exception of the case reported by Teng and Gordon (6), all the intradural-extramedullary lesions were dorsal in location. The majority of reported intradural teratomas were cervical or thoracolumbar.

In our review of reported intradural teratomas, we found no spinal axis congenital anomalies in half of the patients and among the remainder, spina bifida was the most common associated congenital, spinal axis anomaly. Intradural germinal cell tumours have been reported in association with a variety of spinal axis congenital anomalies with spina bifida occulta at the level of the lesion the most frequent. Both diastometamyelia and diplomyelia have been reported in association with these tumours (3,6).

The question of whether this tumour might be an enterogenous cyst was also raised. However, most authors reserved the term "enterogenous cyst" for growth arising exclusively from the foregut, i.e., from the endodermal layer. Therefore, the dorsal location of this tumour in contradistinction to the ventral origin of enterogeneous cyst excludes this by definition. In other words, teratomas are regarded as true maldevelopmental lesions, whereas enterogeneous cysts are considered simple developmental tumours, resulting from the misplacement of normally developing somatic cells (1). Teratomas grow by progressive mitotic division of the cells and whether benign or malignant they are neoplasms.

Congenital dermal sinuses are midline, epithelial-lined tubes that extend inward from the skin surface to a variable depth. They manifest clinically as a pinpoint ostium in the dorsal midline and a small tuft of sparse, wiry hairs frequently protrudes from the ostium. In Wright's collected series (7) of 127 midline dermal sinuses, 1 was sacrococcygeal, 72 were lumbosacral, 12 were thoracic, 2 were cervical, 30 were occipital and 10 were at diverse loci (mostly ventral to the skull base and spinal column). One half to two-thirds of dermal sinuses extend intraspinally. Approximately 60% terminate in deep (epi)dermoid cysts, but in teratomas it is extremely rare. Recently Cybulski (1984) reported an intramedullary cystic teratoid tumour of the cervical spinal cord in association with a teratoma of the ovary (1).

Embryologically, the dermal sinus represents a segmental adhesion between the superficial ectoderm, and the spinal cord. Because the spinal cord becomes buried beneath the surface and then ascends to its adult location as the embryo matures, the segmental dermal sinus extends deeply, becomes elongated and ascends cephalically as it passes from the skin surface to the spinal cord, as in our case, from C5 to C4. The dermal sinus may extend deeply through the median raphe or a bifid spinous process as it did in our case through C4. It may merge with the dura and stop. If it penetrates the dura, it may terminate in the subarachnoid space and discharge cerebrospinal fluid or traverse the subarachnoid space to terminate in the conus medullaris, the filum terminale or the cauda equina as a fibrous nodule in the dorsal column of the cord or an (epi) dermoid cyst or, rarely, a teratoma, as in our case.

Sagittal MRI will display the extraspinal portion of the sinus tract but is less useful for displaying the portion of the tract within the spinal canal. It will show any low-lying spinal cord and often associated intraspinal (epi)dermoid tumours or teratomas which appear hyperintense in T2-weighted images and hypointense in T1-weighted images. In our case MRI displayed the extraspinal portion of the sinus tract and the teratoma hypointense on T1-weighted images and the teratoma appeared hyperintense on T2-weighted images. Although benign intradural teratoma of the lumbar spine was studied with MRI, in the review of the literature we did not find any MR-scan of an intradural extramedullary spinal teratoma of the cervical spine with a congenital dermal sinus.

For Correspondence: Ahmet Erdoğan, M.D.,
Ankara University
School of Medicine
Department of Neurosurgery
A.Ü. Tıp Fakültesi
İbni Sina (Avicenna) Hastanesi
06100 Ankara - TÜRKİYE
Fax: 90 4 125 81 66

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