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CASE REPORT

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Split cord malformation and cystic teratoma masquerading as lipomeningomyelocele

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Abstract *Introduction:* Split cord malformations are commonly associated with other tethering lesions such as short, thick filum terminale and intraspinal lipomas. However, the occurrence of split cord malformation with teratoma is rare. Only 16 such cases have been reported in the English literature. *Case report:* A case of split cord malformation with teratoma masquerading as lipomeningomyelocele is reported, and the possible embryogenesis of this lesion is discussed.

Keywords Diastematomyelia · Teratoma · Split cord malformation · Magnetic resonance imaging

Introduction

Split cord malformations may be associated with meningomyelocele, meningocele, dermal sinus tract, meningomyelocele manqué, dermoid cyst, intradural arachnoid cyst and teratoma, in declining order of frequency [4]. The association of split cord malformation with teratoma is rare, only 16 cases having been reported to date. A case of split cord malformation with cystic teratoma masquerading as lipomeningocele is presented and the possible embryogenesis of this lesion is discussed.

Case report

A 25-day-old female baby was brought for neurosurgical consultation because of a swelling that had been present in the thoracic region since birth. Examination revealed a soft swelling 8 cm long and 3 cm wide and covered by normal skin. There was no transillumination. The child had a normal head circumference with lax fontanelles. Functions in the lower extremities functions were normal. Sphincter functions were also normal. A clinical diagnosis of lipomeningomyelocele was made. Plain radiographs revealed spina bifida at the mid-thoracic level. MRI of the spine revealed spina bifida at the mid-thoracic level, with a type II split cord malformation and a meningocele (Figs. 1, 2). The extraspinal compo-

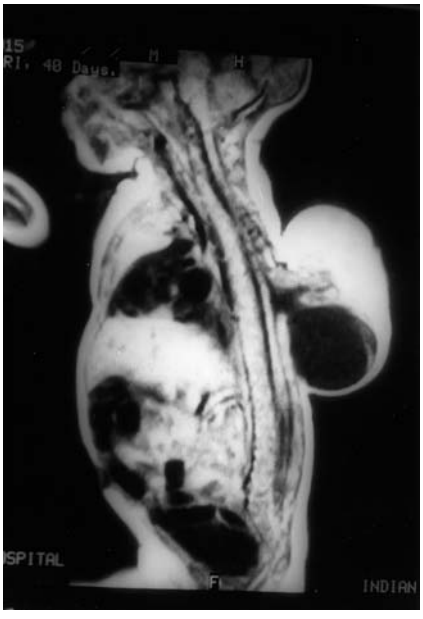

Fig. 1 MRI sagittal sections showing a large, partially cystic lesion in the mid dorsal region with intraspinal extension

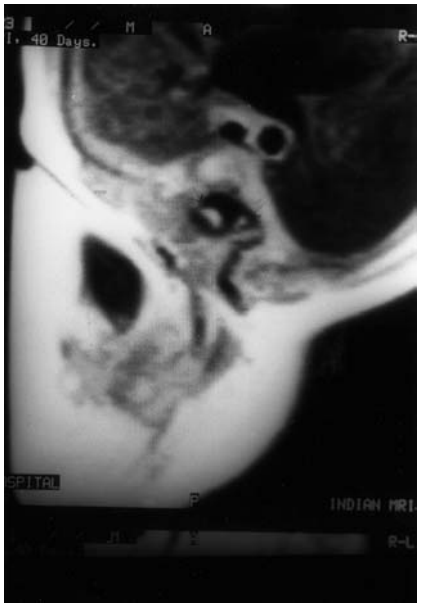

Fig. 2 MRI axial sections showing a split cord malformation with a dorsal lesion having varying signal intensities

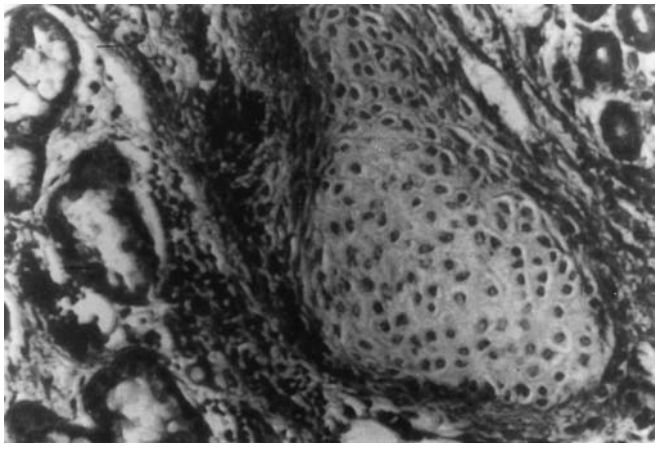

Fig. 3 Photomicrograph showing glandular and cartilaginous structures. (Hematoxylin and eosin; ×240)

nent of the lesion had mixed signal intensities. MRI of the brain revealed tectal beaking with mild tonsillar ectopia. There was no ventriculomegaly. During surgery, a linear incision was made over the swelling. Tissues with evidence of cartilage were removed from the cranial half of the swelling. In the caudal portion of the swelling cystic spaces containing mucinous material interspersed with solid portions were encountered. Dissection led into the extradural compartment, from where the lesion was excised. When the dura was opened a type II split cord malformation was revealed. Postoperatively, the patient's neurological status remained unchanged. Histopathological examination of the lesion exhibited features of cystic teratoma (Fig. 3).

Discussion

The most common location of spinal teratomas the sacrococcygeal region. Teratomas occurring in other regions of the spine are rare [24]. Split cord malformations may be associated with meningocele, meningomyelocele, dermoid cyst, intradural arachnoid cyst, dermal sinus tract and neurenteric cyst [4, 5, 15, 17, 18]. However, association of a split cord malformation with a teratoma is rare. Only 16 such cases have been reported in the English literature (Table 1) [2, 3, 4, 5, 6, 7, 8, 9, 13, 14, 17, 20, 21, 22, 25]. Among the 74 patients with split cord malformations reported by Ersahin et al., 85% had associated lesions [4]. However, there was only 1 with an associated teratoma. Pang reported his results in 39 patients with split cord malformations. Among the 39 patients, 33 had associated tethering lesions. However, none of these 33 patients had a teratoma [19]. Iskander et al. studied 20 patients with meningomyelocele and split cord malformations [7]. They found associated lesions in 12 of the 20 patients. Only 1 of these had a teratoma.

A teratoma occurring in association with split cord malformation may be in the extraspinal, extradural, intradural or intramedullary location. Among the 16 cases of split cord malformations with teratoma reported to date, 9 were intradural, 3 were intramedullary, and the remaining 4 were extradural/extraspinal in location (Table 1). The incidence of teratomas in association with split cord malformations or other forms of occult spinal dysraphism is probably underestimated. Lellouch-Tubiana et al. [11] investigated the histopathology of 234 cases of intraspinal lipomas treated over a 20-year period and found that these "lipomas" may have contained foci of tissues of endodermal, mesodermal and ectodermal origin, suggesting that at least some of them might have a teratomatous component [11].

Teratoma: tumor or malformation?

According to the recent WHO Classification of CNS Tumors, teratomas are tumors that differentiate along ectodermal, endodermal and mesodermal lines (i.e., they recapitulate somatic development from the three embryonic germ layers) [23]. They are further subdivided into mature, immature and malignant types. Mature teratomas are composed exclusively of fully differentiated, 'adult-type' tissue elements that are sometimes arranged in a pattern resembling normal tissue relationships. Mitotic activity is low or absent. Immature teratomas are composed of incompletely differentiated components resembling fetal tissues. Malignant teratoma is the generic designation for the occasional teratomatous neoplasm that contains an additional malignant component in the form of a cancer of conventional somatic type. Such tumors are most often rhabdomyosarcomas or undifferentiated sarcomas.

Table 1 Information reported on published cases of split cord malformation associated with teratoma

Reference	Year	Location	Associated lesions
[13]	1951	Intramedullary	None
[2]	1957	Extramedullary	Meningomyelocele
[21]	1968	Intradural	Meningocele
[8]	1969	Intra- and extradural	Split notochord syndrome
[22]	1969	Intra- and extradural	Meningocele
[25]	1980	Extraspinal	Situs inversus
[25]	1980	Extraspinal	Lumbar spina bifida
[5]	1983	Extradural	Intramedullary epidermoid
[3]	1984	Intramedullary	None
[14]	1984	Intradural	None
[20]	1995	Extradural	Split notochord syndrome
[9]	1998	Intradural	Meningomyelocele
[9]	1998	Intradural	Meningomyelocele
[9]	1998	Intradural	None
[4]	1998	Intradural	None
[6]	1999	Intramedullary	Scoliosis
[17]	1999	Extradural	Meningomyelocele

Recently, Koen et al. [9] reported four cases of intradural spinal teratomas and postulated that teratomas, especially those that occur in relation to spinal dysraphism, may have a dysembryogenic origin, because

1. Several cases of teratomas have been reported in association with dysraphic processes.
2. Most central nervous system teratomas occur in the midline structures and could be derived from the pluripotential cell rests at sites of early neural tube closure.
3. A dysembryogenic origin has been suggested for other neoplasms that typically occur in the midline, e.g., medulloblastoma.

On the basis of these observations, these authors believe that a dysembryogenic process may give rise to intraspinal teratomas.

Currently, the controversy regarding the neoplastic or dysembryogenic origin of teratomas remains unsettled. Germ cell tumors that occur in other regions of the nervous system have a tendency to undergo malignant transformation. However, malignant transformation of a teratoma occurring in association with split cord malformation is rare [13]. Whether this is fortuitous or attributable to the dysembryogenic origin of teratomas that occur in association with split cord malformations in not clear at present.

Embryogenesis of split cord malformation associated with teratoma

The most common location of a spinal teratoma is the sacrococcygeal region. It is believed that these sacrococcygeal teratomas can arise from any one of the following:

1. Totipotential cells derived from the Hensen's node [12, 16]
2. Totipotential cells of the primitive yolk sac [1]
3. Primitive germ cells arrested in midline migration [10]

It is interesting to speculate on the association of split cord malformation with teratomas. This uncommon association can be explained by the currently accepted theory of the embryogenesis of split cord malformations [18, 19]. According to this theory, all split cord malformations arise from one basic ontogenetic error occurring around the time when the primitive neurenteric canal closes. Thus, an accessory neurenteric canal forms through the trilaminar embryonic disc, connecting the yolk sac and the amniotic cavity. This fistula results in the formation of split notochord and split neural plate. As this fistula is lined by the endoderm from the base of the fistula and the mesenchyme condenses around it, the fistula becomes converted into an "endomesenchymal tract." The final evolution will depend on the pattern of healing of this endomesenchymal tract. Persistence of the endodermal elements might result in the formation of neurenteric cysts [15]; persistence of the ectodermal connection of the fistula results in the formation of a congenital dermal sinus. The mesenchymal cells are also known for their pluripotentiality. As the fistula connects all the three germ layers, remnants of all the three germ layers may persist along the fistula and may give rise to teratoma.

Conclusions

Although rare, the entity of teratoma should be entertained in the differential diagnosis when a child presents with a split cord malformation and a skin-covered mass in the back.

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