Magnetic Resonance Imaging findings in a case of intramedullary teratoma of conus medullaris

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Citation

Abstract
A 6 year old female patient presented with complaints of urinary and fecal incontinence. There was also progressive weakness of both the lower limbs. Physical examination revealed significantly reduced motor and sensory functions in both the limbs. Magnetic Resonance Imaging (MRI) revealed presence of a large intramedullary tumor in the conus medullaris causing its marked expansion. Presence of calcification and fat in the tumor was characteristic of teratoma.

Teratomas of the nervous system are rare tumors, the most common site being pineal region. Intraspinal teratomas are rare, usually developed in sacrococcygeal region and often seen in neonates. Intramedullary teratomas are extremely rare and their occurrence in pediatric patients is rarer still. Here we present a case of a large intramedullary teratoma in a pediatric patient.

CASE REPORT
A 6 year old patient presented in the pediatric out patient department with the complaints of urinary and fecal incontinence for the last 3-4 months. Patient also complained of progressive weakness of both the lower limbs. There was history of numbness in lower limbs. Physical examination revealed marked deterioration of both sensory & motor functions of both lower limbs. The deep tendon reflexes were markedly impaired and plantars were upgoing. Urinary bladder was distended. The patient was referred for MRI to rule out any spinal pathology. MR examination of lumbosacral spine was performed on a 1.5T magnet using dedicated coils.

MR images revealed a large well defined lesion replacing the conus medullaris and causing its marked expansion. Associated expansion of the spinal canal was also seen due to mass effect. The lesion showed mixed intensity pattern with a large component appearing hyperintense on both T2WI & T1WI and was following intensity pattern similar to subcutaneous fat suggesting fat component. On fat suppressed images the fat component showed signal suppression further confirming its fatty nature. A round component was seen anteroinferiorly showing hyperintense signal on T2 weighted images(WI) & hypointense on T1WI (Figures 1 and 2) suggesting fluid component. A small nodular area was seen showing hypointense signal on all sequences suggesting calcification. Walls of the cyst were well defined and no evidence any infiltration of surrounding structures was seen. The presence of fat, calcification and fluid in this case are typical of teratoma.
Figure 1
Figure 1 : Sagittal T1 weighted images of the lumbosacral spine revealed a large well defined solid cystic mass in the conus medullaris. The lesion shows predominantly hyperintense signal similar to subcutaneous fat. A smaller round component in its lower part anteriorly showed hypointense signal. Another smaller nodular area is seen in central part in the second scan appearing hypointense. Spinal canal is markedly dilated by the mass. There is evidence of scalloping on the posterior surface of the vertebral bodies.

Figure 2
Figure 2 : Sagittal T2 weighted images revealed the lesion predominantly to be hyperintense as was seen on T1WI suggesting fat component. The component in its anteroinferior part appears hyperintense that showed hypointense signal on T1WI suggesting fluid/soft tissue component. The central nodular area appear hypointense similar to T1WI suggesting calcification.

DISCUSSION
Intramedullary spinal teratomas are extremely rare representing 0.2-0.5% of all spinal cord tumors. The most common site is thoracolumbar region. There is some confusion regarding the nomenclature of these tumors with many terms in vogue. Some of the terms used commonly are teratoma, teratomatous cyst, cystic teratoma mature & immature teratoma etc. Besides this these lesions has been confused with neurenteric cysts. Willis defined the teratomas as malformations composed of multiple tissues foreign to their localization and lacking organ specificity. They arise from the detachment and proliferation of cells that escape primary inducer control during early embryonic development and grow elsewhere chaotically. It must be noted that there is no mention that all the three layers must be represented. Although a teratoma represents a neoplasm composed of all three germ cell layers, the presence of only two layers does not rule out the diagnosis.
Like all other extragonadal germ cell tumors, central nervous system teratomas tend to occur in the midline with the pineal region being most common followed by other rare sites like suprasellar region, cerebral hemispheres and posterior fossa. Intraspinal teratomas very rare. Some authors commented that the intramedullary variety of teratoma is extremely rare, having been reported only seven times during the last 30 years. Among the spinal teratomas dorsal or dorsolateral location is usual and the mass may be extra- or intramedullary.

Diagnosis of the intramedullary teratomas by the imaging is straightforward as in our case. Some authors have advised Computed Tomography (CT) for the preoperative diagnosis as it can show calcifications, fat and soft tissue densities. However with the technological advances MRI has become the imaging investigation of choice for preoperative diagnosis for teratomas as for other intraspinal tumors. MRI can show different tissues with the help of tissue specific sequences for example fat can be recognized on simultaneous non-fat saturated and fat saturated sequences and calcifications can be better detected by gradient echo sequences. Presence of enhancing soft tissue components is better depicted by MRI. Moreover the exact location, morphology and its relation to surrounding structures is best demonstrated by the MRI. However calcifications some times can be missed on MRI but detected on CT. Thus the presence of a cystic lesion showing fluid, fat and calcifications are diagnostic of teratoma.

Total surgical removal should be the aim of the surgery, as incomplete removal of the tumor can lead to re-growth of remaining endodermal or ectodermal elements. If completely and successfully excised, mature teratomas have a good prognosis with long term disease free survival. Such a favorable course may be correlated to the degree of differentiation. Poeze et al explained that treatment of these tumors is surgical, especially since an exact diagnosis as benign or malignant cannot be made preoperatively.

References
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