Case Report

Intraspinal Teratomas- a Report of two cases and Review of Literature

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ABSTRACT

Intraspinal teratomas are rare congenital inclusion tumors. We report here two cases of spinal mature teratomas in adult patients, at different locations in the spinal canal. In one patient the tumor was intramedullary, located at conus medullaris and not associated with spinal dysraphism, while in the other, tumor was at the lumbosacral level intradurally, associated with hemivertebral defect of S1-2 level. These tumors are rarely diagnosed before surgical intervention. The clinical features, magnetic resonance imaging characteristics and surgical treatment of such rare tumors are discussed and the relevant literature reviewed.

KEY WORDS: Teratomas, spinal cord.

INTRODUCTION

Intraspinal teratomas are rare lesions with an average incidence of 0.2 to 0.5% of all spinal tumours and are often found in association with spinal dysraphism.[1 ,8] Thoraco-lumbar region is the most commonly affected site in adults. They are thought to arise from misplaced pleuripotent cells in midline during early embryogenesis.[2] Teratomas may be mature, immature and malignant according to their histological characteristics.[6, 9]

CASE-1

Twenty six year old male presented with a two year history of paresthesias of both lower limbs along the posterior aspect below the buttocks and the thighs. There was no history of limb weakness or sphincter disturbance. The symptoms were slowly progressive. General physical examination was unremarkable. There were no cutaneous markers suggestive of spinal dysraphism. He had bilateral hyperreflexia in lower extremities. Sensory examination and sphincter control was normal. MRI showed large SOL, well defined oblong shaped cystic lesion with heterogenus signals in the conus opposite D12-L1 region. [Figure 1] Intraoperatively the lesion was located in the conus and pushing all the roots to one side. There was an abscess at the rostral end of the lesion within the conus. Rest of the lesion contained cholesterol flakes and total excision done with microscopic dissection. Histopathology showed mature tridermal differentiation with admixture of stratified squamous epithelium, dermal appendages and immature cartilage. Post operatively he developed urinary retention which also improved partially at 3 months follow-up. No recurrence is seen after 1 year.

CASE-2

Twenty eight year old male presented with severe radicular pain of both lower limbs with progressive weakness of both lower limbs of one year duration. There was history of urinary incontinence. On examination he had 2/5 power distally in both lower limbs. Ankle jerks were absent bilaterally. There was 50% reduction of sensory loss in L5, S1-3 dermatomes bilaterally. MRI revealed evidence of well defined altered signal intensity lesion noted at distal spinal cord (conus medullaris) which is intradural and extramedullary and is heterogenous with few areas of hyper intense areas on T2WI.[figure 2] Intra operatively there was hemivertebral defect of both L5&S1. Tumour was intradural. A large lobulated mass with a terminal lipoma was found. Tumor was adherent to thick film which was excised. Cyst contents were pearly white with tufts of hair. Near total excision of the tumor could be achieved with microscopic dissection. Histopathology showed cyst lined by stratified squamous epithelium with adnexa, sebaceous glands and keratin material. Subepithelium showed extensive areas of fibrocollagenous tissue, adipose tissue and foci of glial tissue, suggestive of mature teratoma.[figure 3] Post operatively patient progressed well, regained his power to near normal. At the end of 1 year bladder dysfunction persisted.

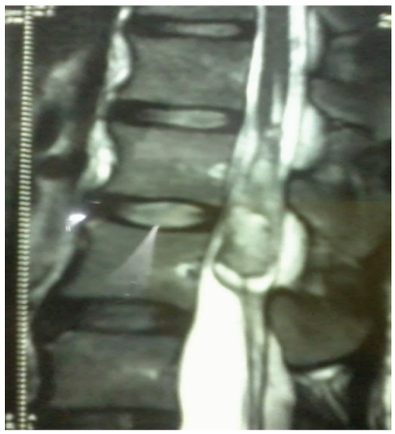

Figure 1
Well defined oblong shaped cystic lesion with heterogeneous signals in the conus opposite D12-L1 region

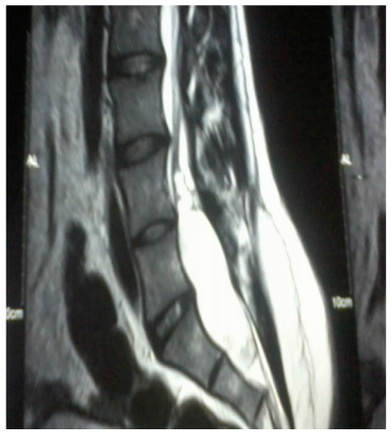

Figure 2
Well defined altered signal intensity lesion noted at distal spinal cord (conus medullaris) which is intradural and extramedullary and is heterogeneous with few areas of hyper intense areas on T2WI.

Figure 3
High power magnification, 10 X showing stratified squamous epithelium, keratin, and mucinous glands.

DISCUSSION

Teratomas are rare intraspinal tumors accounting for less than 0.5% of all intra spinal tumours. They are found in extradural, intradural and intramedullary locations. They have been reported in association with spinal dysraphism especially in children.[8] Intramedullary location was commonly observed in pediatric population specially when associated with spinal dysraphism [3]. Willis defined the teratomas as malformations comprising of tissues foreign to their localization and lacking organ specificity. Teratomas can occur at many locations throughout the body. In the CNS, teratomas constitute about 0.5% of mass lesions and tend to occur in young children in the midline, excluding sacrococcygeal teratomas in neonates which accounts for 40% of all teratomas. Intra spinal teratomas are rare tumors [1]. Intramedullary teratomas are still rare. Only 30 cases of intramedullary teratomas were reported so far in literature.

Most frequent site of the tumor was conus medullaris and lumbo sacral region followed by thoracic spinal cord in dorsal and dorso-lateral locations. Teratomas in cervical region are very rare and only four cases of intramedullary teratomas were reported so far [8].

There is no consensus regarding the nomenclature used for this group of lesions. Teratomas, teratomatous cysts and teratoid tumors are the alternative terms used commonly in literature. The term Teratoid is used when derivatives from only two layers is present, the bigerminal teratoma.[6, 9] Several hypotheses have been proposed for the origin of spinal teratomas. The most accepted of which is based upon the fact that misplacement of primordial germ cells into dorsal midline structures during their normal migration from primitive yolksac to gonadal ridges can develop into germinomas and teratomas. In addition their well known association with dysraphic anomalies of spine suggests that a developmental fault arose during embryogenesis in the pleuripotent embryonic caudal mesenchyme giving rise to teratomas related to dysfunction of several factors that probably involve genetic and cellular inductive interactions. Though there is no specific age prevalence reported, these tumours are common between the age group of 5yrs to 30yr,

often with a long history of symptomatology ranging from 6m-2yrs. Males were found to be more commonly involved.

Clinical presentation are usually neurological, like any other SOL such as progressive paraperesis, paresthesias of one or both lowerlimbs and urological disturbances according to the site of the tumour location.[7] Neurological examination may reveal presence of radiculopathy or myelopathy or both with presence of associated anomalies of spinal dysraphism like hemivertebra, tethredcord or a terminal lipoma etc [8, 10].

MRI is the gold standard of investigation for pre operative diagnosis of teratomas as for other intraspinal mass lesions [4]. The presence of cystic lesion showing fat, fluid and calcifications are characteristic of teratomas [6, 7] . MRI can show different tissues and the finding of mixed high and low signal intensities reflect cystic and solid components of the tumour

Histologically teratomas contain derivatives from all three embryonic layers. They are termed mature or immature depending on the differentiation of elements [1]. Besides the usual admixture of skin and adnexal elements muscle, fat, neural tissue, bone and cartilage, intestinal and respiratory elements have been reported [5]. The presence of two layered elements does not rule out the diagnosis of teratoma.

Laminectomy/laminotomy for children and radical excision of the cyst by microsurgical techniques without endangering neural structures offers good outcome for mature teratomas. When there is no definite plane between the tumour and cord structures near total excision leaving some areas adherent to neural structures can be left over. Though these are benign tumours histologically, research shows 10% recurrence rates [6, 7]. Subtotal excision offers a long period of symptom free life. Recurrence is commonly observed when the cysts contain immature elements in the cyst and radiotherapy should be considered for immature teratomas.

CONCLUSION

Intraspinal teratoma is a rare congenital inclusion tumor. There are no specific clinical or radiological features for the pre operative diagnosis of these lesions. The presence of spinal dysraphic defects should raise the index of suspicion for the presence of an associated teratoma. MR images are helpful in delineating the solid /cystic components of the tumor. Diagnosis is based on intra operative findings and thorough histopathological examination of the entire specimen for the presence of derivative elements of all three germinal layers. Total excision with microsurgical techniques is the primary treatment modality. Gross total removal offers good prognosis and long term symptom free life.

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