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Case Report

CASE REPORT

Diastematomyelia due to Spinal intradural Extramedullary Teratoma in an Adult

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ABSTRACT

Teratomas are rarely found in the Central Nervous System and are approximately 2% of all other Teratomas. Spinal Teratomas are extremely rare as compared to the teratomas that are found intracranially and are almost 0.1–0.5% of all tumors of spinal cord. These tumors are even more infrequent in adults. Diastematomyelia, also known as split cord malformation causes complete or incomplete sagittal division of the neural axis and duplicates it. This condition is mostly accompanied by various malformations and in rare cases it is associated with intradural spinal teratoma. We report a case of 39 years old male who presented with complaints of severe pain in the left leg and weakness with power 2/5 in left leg and urinary obstruction. On evaluation, he was diagnosed as having Diastematomyelia due to Spinal intradural Extramedullary Teratoma.

Key Words: Diastematomyelia, Spinal intradural Teratoma, Split cord malformation

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Introduction

Diastematomyelia, also known as split cord malformation causes complete or incomplete sagittal division of the neural axis and duplicates it.[1] This condition is mostly accompanied by various malformations including skeletal and vertebral anomalies, meningocele, myelomeningocele, cutaneous changes, hydromyelia, hydrocephalus, Klippel–Feil syndrome, Arnold–Chiari malformation and in rare cases it is associated with intradural spinal teratoma.[2] Intradural spinal teratomas are very rare dysembriogenetic tumors [3] and are less common in adults.[4] The first case of spinal teratoma was reported by Virchow in 1863.[5]

Among total spinal tumors, the incidence of intradural extramedullary spinal teratoma (IEST) is only 0.15-0.18%.[6] Whereas in infants and children the incidence of spinal teratomas is comparatively higher and is approximately 5–10% of all spinal tumors.[7] Furthermore, the ratio among male to female is 3:1.6. Intradural extramedullary spinal teratoma is mostly located in dorsolumber region and is associated with conditions like spinal dysraphism prior spinal surgeries and lumbar puncture (LP) .[8] For IEST, magnetic resonance imaging (MRI) is the diagnostic modality of choice, whereas total surgical excision is the treatment modality of choice for it.[9] Teratomas are classified into mature, immature and malignant type, on the basis of histological characteristics and degree of differentiation.[10] These tumors typically, but not always are composed of tissue derived from three germ cell layers' ectoderm, mesoderm, and endoderm.

In Infancy, these tumors usually arise from sacrococcygeal region although it can arise from anywhere throughout the spinal canal in cases of adults, but in reported cases, there is predominance of thoracic and lumbar region and the most frequently involved site is conus medullaris.[11].

Case Report

A 39 years old male presented via OPD as a case of low back pain and left sided lower limb weakness since his

childhood with sudden onset of urinary symptoms ranging from dysuria to obstruction in the last 15 days. Patient had multiple episodes of pain in his childhood, which improved with physiotherapy and medication. Now the patient presented with severe pain in the left leg and weakness with power 2/5 in left leg and urinary obstruction. On examination, tuft of hairs in the lumbar region along with a dermal sinus without discharge was seen. His left leg had gross muscle atrophy in calf muscles measuring 3cm less than the right calf. Patient had painful dorsiflexion, grade 4/5. Power in the left lower limb was 4/5 and 5/5 in the right lower limb and the patient also had bilateral high arched feet. He was advised X-Ray Lumbosacral spine which showed spina bifida in the lower lumbar region. MRI Lumbosacral spine with contrast was done which showed features of Diastometomyelia due to intradural space occupying lesion (SOL), most likely dermoid cyst (Figure 1). Patient was counseled regarding the condition and surgical treatment. Surgery was performed and excision of intradural mass was done and sent for histopathological examination.

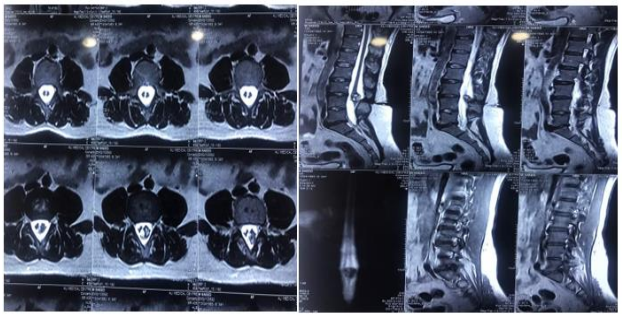

Figure 1: MRI showing Diastematomyelia due to Spinal intradural Extramedullary Teratoma.

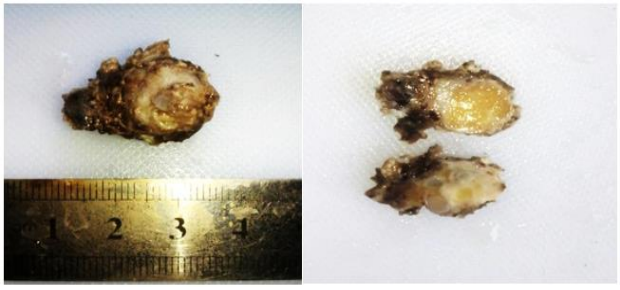

Figure 2: Gross appearance of Spinal Teratoma showing cystic and solid areas

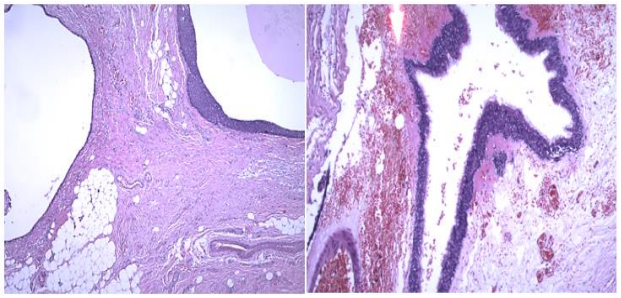

Figure 3: Histopathological examination showing various mature derivatives of germ layers fibroadipose tissue and small cystic areas lined by respiratory and stratified squamous epithelium. H&E, 400X

On gross examination, the specimen comprised of a cystic mass, its cut surface showed cystic and yellowish white solid areas (Figure 2). Histopathological examination revealed various mature derivatives of germ layers, comprising of mature neural tissue, fibroadipose tissue, mature bone and small cystic areas lined by respiratory and stratified squamous epithelium. Focal areas of calcification were also seen, whereas no immature derivatives were seen (figure 3). Patient recovery was unremarkable except power of left lower limb of 3/5.

Discussion

Teratomas are rarely found in the Central Nervous System (CNS) and are approximately 2% of all other Teratomas.[12] Most of the CNS teratomas are seen intracranially and mainly involve the midline structures and are mostly seen in the pineal region, some in suprasellar compartment and rarely found in fourth ventricle, basal ganglia, posterior fossa and cerebral hemispheres.[13] Spinal Teratomas are extremely rare as compared to the teratomas that are found intracranially and are almost 0.1–0.5% of all tumors of spinal cord.[12] These tumors are even more infrequent in adults.[14] In one study that was conducted by Slooth et al. in 1964, out of 1322 spinal cord tumors only two were of spinal teratomas. In another series of neurosurgical biopsies conducted by Al-Sarraj et al, in a total period of 15 years there were 25,000 total cases, out of which only seven were of spinal teratomas.[15] Clinically, these spinal teratomas of adults present variably depending upon the location. Symptoms of the patient depend on whether the

tumor is present in the cervical region, thoracic or in lumber region. The presentation also varies in cases of intramedullary, extramedullary, intradural or extradural Teratomas. Poeze et al. reviewed intramedullary teratomas (31 cases) out of which 22(71%) cases presented with motor dysfunction and that was the commonest complaint in those patients.[15] The other complaints were of disturbance in reflexes seen in 16 (52%), and sensory changes seen in 14 (45%) cases. Some cases (11, 35%) also presented with pain and urinary disturbance. Other complaints that were only seen in one or two patients included disturbance in defecation, meningismus, visible tumor and sexual disturbance. In a study by Tarantino et al, pain was the commonest symptom (47.7%) observed in 107 cases that presented with intradural, extramedullary tumors[16] and other less common symptoms were of sensory deficits (15%) followed by motor deficits in 12% cases, sensorimotor deficit in 19.6% cases and sphincter dysfunction in 3.7% of cases.[15]

Diastematomyelia also known as split cord malformation (SCM) is accompanied by intradural Teratoma in rare cases. SCM is of two types. In case of Type I SCM, there are two hemicords each having its separate dural sheath and an osteocartilaginous septum separates the two hemicords. In Type II SCMs there are two hemicords but both have same dural envelope and a fibrous septum separates them.[2] In our case, the patient was shown to harbor a type II lesion. Caruso, et al,[3] reviewed 33 cases of Intramedullary Spinal Teratoma out of which two cases presented with Diastematomyelia. Hader, et al.,[4] reviewed 12 cases that presented simultaneously with intradural spinal teratoma and SCM but out of these only two cases were found in adults.[17] If an adult presents with spinal teratoma, radiological evaluation of whole spinal canal should be done to rule out dysraphic lesions in any spinal area. The most common symptom seen in adults that present with occult spinal dysraphism is pain which may be accompanied by neurological deficit. When Teratoma and SCM occur simultaneously, it usually increases neurological problems and may lead to a complex surgery.[1] Regarding the origin of intraspinal teratoma there are two different theories[18], one theory is Dysembryogenic theory and the other is misplaced germ cell theory. In case of Dysembryogenic theory, spinal teratomas originate from pluripotent cells that differentiate chaotically and this occurs in a developmental environment that is locally disturbed. When this occurs in a caudal cell mass or primitive streak, it develops in to a spinal teratoma. The other theory is the misplaced germ cell theory in which some of the pluripotent primordial germ cells from the neural tube, while migrating from yolk sac to gonad get misplaced and in this way develop into spinal teratoma. This germ cell theory is more likely to be responsible for adult intraspinal teratomas and which normally do not lead to significant dysraphism.[18]

The gold standard technique used for the diagnosis of Spinal teratomas is MRI by which the location and the extent of involvement is revealed. The solid and cystic areas are also identified by this technique. But the definitive diagnosis can only be made by histopathological examination, which clearly confirms the presence of three germinal layers or two germinal layers (ectoderm, mesoderm, and endoderm).[11] It is suggested that the tumor should be resected as much as possible keeping in mind that the surrounding neural tissue should be completely preserved. It should also be noted that while performing surgery, the content of cyst should never be spilled into the intradural space because it can cause chemical meningitis.[19] Regarding the treatment of spinal teratomas, the role of chemotherapy and radiotherapy for treating the residual portion of mature teratoma is not clearly evident, mainly due to its slow growth and rarity. However, in case of Immature Teratoma (malignant) it is controversial to use these techniques.[6] In this case, only surgical excision was done and tissue was totally removed. There was no need of adjuvant therapy because on histopathology this tumor was diagnosed as non-malignant mature cystic teratoma. Follow up of the patient is recommended by performing serial MRI especially if the patient develops new neurological signs and symptoms, if the diagnosis is immature teratoma or if adjuvant therapy is used.[9]

Conclusion

Only a few cases of Intraspinal teratomas are reported in the literature and it is even more rare in adults. The presence of Diastematomyelia along with spinal intradural teratoma in adults is very rare. Among the imaging modalities, MRI is very useful for identifying the location,

extent and determining solid or cystic composition. However, the final diagnosis depends on the histopathological examination. The main treatment of choice for these tumors is aggressive surgical resection but in cases of adhesion to the neurological components, subtotal resection is more appropriate in order to prevent from complications.

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