Original Research Paper

Volume - 10 | Issue - 7 | July - 2020 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Neurosurgery

EWING SARCOMA OF CAUDA-EQUINA REGION: CASE SERIES WITH REVIEW OF LITERATURE

Dr Anand Sharma Assistant professor Department of NeurosurgeryGajra Raja medical College

Dr Parth Lalcheta* Mch Neurosurgery Resident Department of Neurosurgery Gajra Raja medical College *Corresponding Author

Dr Avdhesh Shukla Associate professor Department of Neurosurgery Gajra Raja medical College

Dr S N iyengar Professor and Head Department of Neurosurgery Gajra Raja medical College

ABSTRACT Ewing's sarcoma family of tumors (ESFT) is a group of small round cell tumors of neuroectodermal origin. It usually affects children and adolescents. Primary Ewing's sarcoma of the spine is rare. It occurs predominantly in the paravertebral regions and epidural spaces, and the involvement of the intradural space is decidedly rare. The treatment consists of wide surgical resection within safe limits, followed by chemoradiation and local irradiation. In this case series we reported cases of cauda-equina Ewing's sarcoma with review of literature.

KEYWORDS : Ewing Sarcoma, Cauda-equina region

INTRODUCTION:

Ewing's sarcoma family of tumors (ESFT) is a group of small round cell tumors of neuroectodermal origin and includes Ewing's sarcoma of the bone, extra skeletal Ewing's sarcoma, primitive neuroectodermal tumor, and Askin tumor. Primary Ewing's sarcoma of the spine is rare. It usually affects children and adolescents. It occurs predominantly in the paravertebral regions and epidural spaces, and the involvement of the intradural space is decidedly rare. They present with local pain, gait disturbance, motor deficit, or sphincter dysfunction. Peripheral PNET (pNET) of spine refers to all those tumors arising from the surrounding soft tissues vertebra or spinal nerve roots. The mean period of postoperative survival ranges from 18 to 20 months. Nearly one third of the spinal PNET present with Cauda equine syndrome [1]. In this case series we reported cases of cauda-equina Ewing's sarcoma with review of literature.

Case report: 1-

45-year male reported to neurosurgery OPD with low back pain with radiation to both lower limbs of 2-month duration. Neurological examination revealed perianal sensory loss, no other neurological deficit noted. MRI Lumbosacral region revealed intradural lesion at L5-S1 location, hypointense on TIW images and heterogenous hyperintense of T2W images. On post contrast images there is heterogenous enhancement. CXR, CT Brain and USG abdomen were normal. Provisional diagnosis of ependymoma/nerve sheath tumor was made. After discussion of risk and benefit surgery was planned. L5, S1 laminectomy with microscopic total excision of mass was done; Intraoperatively tumor was dark red in color, highly vascular, nonsuckable, intermingled with nerve roots. Postoperative course was uneventful, without additional neurological deficit. Histopathological examination revealed monomorphic tumor cells arranged around blood vessels, forming pseudorossests likely small cell tumor. Immunohistochemistry were positive for CK, CD 99, CD56, and FL1 S100 and negative for LCA, Desmin, Synaptophysin, and chromogranin. Diagnosis of Ewing's sarcoma was confirmed. Patient sent for adjuvant therapy (Figure 1-2). Patient was followed for two year had multiple recurrences over lumber and dorsal region.

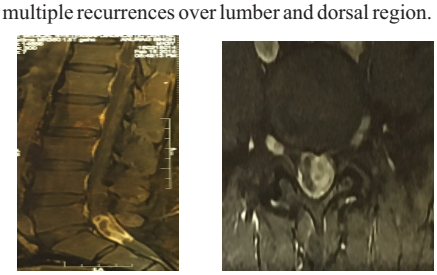

Figure 1: A) Saggital T1 Contrast B) Axial T1 Contrast

Case report 2: 12 year female presented to neurosurgery OPD with severe low back pain radiating towards right lower limb since 15 days duration. Neurological examination revealed no deficit. MRI whole spine revealed extradural lesion at L4, L5 region, and hypointense on T1 and hyperintense on T2 image. On post contrast images there is heterogenous enhancement. USG abdomen revealed ovarian cyst, CEA was normal. Contrast CT thorax and abdomen did not revealed any additional diagnosis except serous ovarian cyst. Provisional diagnosis of primary bony lesion or suspected metastasis was made. After discussion of risk and benefit surgery was planned. L4, L5 laminectomy with completed excision of mass with decompression of exiting nerve roots was done. Intraoperatively tumor was grayish white in color, breaching in outer laminar surface was present but there was no breach in inner table. Moderately vascular, not suckable, intermingled with nerve roots. Postoperative course was uneventful, without additional neurological deficit. Histopathological examination revealed monomorphic tumor cells arranged around blood vessels, forming pseudorossests likely small cell tumor. Immunohistochemistry were reactive for CK, CD 99, and GATA-3, negative for LCA, Desmin, Synaptophysin, and chromogranin. Diagnosis of Ewing's sarcoma was confirmed. Patient sent for adjuvant therapy.

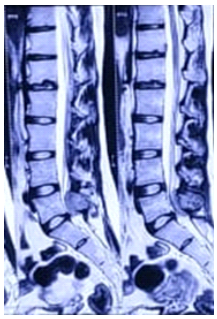

Figure 2: A) Extradural mass over L5-S1 region, hypointense on T2

DISCUSSION

Ewing`s sarcoma and primitive peripheral neuroectodermal tumor (PNET) are round cell sarcomas that belong to the ES family. They varied degrees of neuroectodermal differentiation and represented two end of spectrum of same entity. Most of the Ewing's sarcoma occur in the long bones, pelvis, or ribs [2], but rarely may have an extraskeletal origin that named extra skeletal Ewing sarcoma. EES has similar histology to skeletal Ewing sarcoma, which commonly affects the epidural spaces and paravertebral regions. The involvement of the intradural space is rare, and only 15 cases of primary intradural extramedullary EES have been previously reported [3].

Definitive diagnosis of extra skeletal Ewing sarcoma relies on pathological assessment, and molecular or cytogenetic analysis of the trans location t (11; 22) (q24; q12) has been recognized as the diagnostic gold standard [4]. Pathologically, the differential diagnosis of

EES includes all other small round blue cell tumors: lymphoblastic lymphoma, desmoplastic small round cell tumor, rhabdomyosarcoma, neuroblastoma, primary central nervous system primitive neuroectodermal tumors, and small cell carcinoma. Immunohistochemical, neuroblastoma would be positive for CD56, and a monoclonal lymphoid population would support the diagnosis of lymphoma. The ependymal true rosettes or pseudorossests and a papillary arrangement of cuboidal or columnar tumor cells are morphological characteristic of ependymoma.[4,5] Moreover, both EES and primary central nervous system primitive neuroectodermal tumors are designated PNET and stain for neuroendocrine markers. While vimentin, S100, CD99/MIC2 (in 84% to 100% of cases) as well as neuroendocrine markers including CD56 and Synaptophysin may or may not be helpful in distinguishing these entities, the reverse transcriptase polymerase chain reaction (RT-PCR) for the fusion gene EWSR1-FTI1 is diagnostic. Additional markers, including those derived from muscle (myogenin, muscle specific actin, MyoD1) and the hematolymphoid system (CD3, CD34, TdT), are strategic in ruling out other small blue cell tumors and ultimately help confirm the diagnosis of EES.

Distant metastasis is seen in 38 % cases commonest being the lungs. They are CD 99 positive and associated with t (11, 22) translocation[6].

The treatment consists of wide surgical resection within safe limits, followed by chemoradiation and local irradiation. In the management of Ewing's sarcoma, com- plete resection of the primary lesion is associated with superior outcomes.[7] It is noteworthy that the surgical approach toward other diagnostic possibilities for similar appearing intraspinal lesions also requires an attempt at complete resection: meningioma, schwannoma, myxopapillary ependymoma, paragangliomas, hemangioblastoma, and metastasis.[7] Therefore, surgical planning for a gross total resection, where possible, when such lesions are encountered, may improve outcomes for the rare patient who presents with EES involving the intradural extramedullary spinal canal.

This may lead to improvement of the prognosis[7]. Ewing sarcoma is an aggressive type of tumors with a high incidence of recurrence and metastasis[9], so follow up should be undertaken routinely in order to find the recurrence or metastasis as soon as possible. And it is necessary to restart chemotherapy and radiotherapy in the patients with recurrence, for it may help to control the disease effectively[7].

REFERENCES

[1] Jithendra K S,rachna S ,karak AK ,Thavaraj V , Kabra SK. Primitive Neuroectodermal Tumor presenting as Cauda equine syndrome .Indian J Med and Paediatr Oncol 2007;28:34-37

[2]- Salunke PS, Gupta K, Malik V, Kumar N, Henke LE, Cai C, Chen WS, Pfeifer JD. Primary Ewing's sarcoma of cranial bones: analysis of ten patients. Acta Neurochir (Wien) 2011; 153: 1477-85.

[3] Uesaka T, Amano T, Inamura T, Ikezaki K, Inoha S, Takamatsu M, Iwaki T, Fukui M. Intradural, extramedullary spinal Ewing's sarcoma in childhood. J Clin Neurosci 2003; 10: 122-5.

[4] Song X, Choi J, Rao C, Nallu S, Nicastri AD. Primary Ewing sarcoma of lumbar spine with massive intraspinal extension. Pediatr Neurol 2008; 38: 58-60.

[5] Mobley BC, Roulston D, Shah GV, Bijwaard KE, McKeever PE. Peripheral primitive neuroectodermal tumor/Ewing's sarcoma of the craniospinal vault: case reports and review. Hum Pathol 2006; 37: 845-53.

[6] Saeedinia S ,Nouri M ,Alimohammadi M ,Moradi H ,Amjirjamshidi A Primary spinal extradural Ewings sarcoma(PNET);Report of a case and meta analysis of the reported cases in the literature .Surg Neurol Int 2012 :3:55

[7] Haresh KP, Chinikkatti SK, Prabhakar R, Rishi A, Rath GK, Sharma DN, Julka PK. A rare case of intradural extramedullary Ewing's sarcoma with skip metastasis in the spine. Spinal Cord 2008; 46: 582-4.

[8] Mardekian SK, Gandhe A, Miettinen M, Pack S, Curtis MT, Abdullaev Z. Two Cases of Spinal, Extraosseous, Intradural Ewing's sarcoma/Peripheral Neuroectodermal Tumor: Radi-ologic, Pathologic, and Molecular Analysis. J Clin Imaging Sci 2014; 4: 6.

[9] Arpaci E, Yetisyigit T, Seker M, Uncu D, Uyeturk U, Oksuzoglu B, Demirci U, Coskun U, Kucukoner M, Isıkdogan A, Inanc M, Alkis N, Ozkan M. Prognostic factors and clinical outcome of patients with Ewing's sarcoma family of tumors in adults: multicentric study of the Anatolian Society of Medical Oncology. Med Oncol 2013; 30: 469.