Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: www.saspublishers.com

OPEN ACCESS

Pathology

Primary Malignant Schwannoma of the Uterine Cervix: Unsual Site Posing a Diagnostic Challenge

Monal Trisal, Safia Rana*, Beenish Sultan, Sujata Jetley

Department of Pathology and Blood Bank, Hamdard Institute of Medical Sciences and Research New Delhi India

DOI: 10.36347/sjams.2019.v07i10.041 | **Received:** 05.06.2019 | **Accepted:** 13.06.2019 | **Published:** 29.10.2019

***Corresponding author:** Safia Rana

Abstract **Case Report**

Uterine Sarcomas comprise of less than 1% of the malignancies of the uterine cervix. The most frequently encountered sarcoma in cervix is Rhabdomyosarcoma. Malignant peripheral nerve sheath tumor (MPNST) is very rarely reported in female genital tract. In this paper we present a 22-year young girl with MPNST of the uterine cervix.

Keywords: Malignant peripheral nerve sheath tumor, uterine cervix, malignant schwannoma.

INTRODUCTION

Primary tumors of the uterine cervix are mostly squamous cell carcinoma, followed by adenocarcinoma. Other malignancies arising from the uterine cervix are small cell carcinoma, adenoid cystic carcinoma; lymphoma and melanoma are very rare. Primary pure sarcomas account for less than 1% of malignancies of the uterine cervix [1]. Among them, the ones most frequently reported are the rhabdomyosarcoma. Malignant peripheral nerve sheath tumor (MPNST) is very rare. Malignant peripheral nerve sheath tumors (MPNST) constitute about 10% of soft tissue sarcomas, most arising in association with major nerve trunks, including the sciatic nerve, brachial plexus, and sacral plexus. The most common anatomical sites include the upper and lower extremities and trunk and less commonly the head and neck region. Very few cases have been reported in the female genital tract [2] among patients with Neurofibromatosis-1 (NF-1), the ultimate risk of developing MPNST is approximately 2–4%. About 40–50% of MPNSTs are associated with a family history of NF-1[3].

To our knowledge, only nine cases of MPNST of the cervix had been reported previously and none of them had evidence of NF syndromes. In this paper we present a 22-year-old young girl with MPNST of the uterine cervix.

CASE PRESENTATION

A 22-year-old young unmarried girl came to gynaecological opd of our hospital with complaints of bleeding per vagina with whitish discharge and irregular cycles for one year.

She complained of fever on and off and pain in abdomen for two months. On physical examination, abdomen was soft with mild tenderness and a 14 weeks size uterus was palpable and per vagina bleeding with a large cervical mass was felt anteriorly on retracting the labia minora.

On USG, a heterogenous SOL measuring 95x93com contiguous with cervix was seen suggestive of ? Blood ligament fibroid? cervical fibroid. Abdominopelvic MRI scan was performed. MRI scan revealed a large cervical mass exerting pressure on the bladder and rectum. A well-defined lobulated mass in the pelvis with hyperintense signal on IR and hypointense signal on T1 W images (Figure 1,2).

Lesion was seen causing fraying of cervical lips with bulk of lesion filling the recto-uterine pouch, posteriorly abutting the rectosigmoid and anteriorly indenting on the urinary bladder. Interface with the surrounding structures were well maintained. Vagina was not well visualised separately. The bilateral ovaries and tubes were unremarkable. Intraoperative consultation was carried and a diagnosis of Malignant

Mesenchymal Tumor with large areas of necrosis was rendered (Figure 3).

On gross, the mass was received in form of multiple irregular grey brown to grey tan soft tissue masses weighing 500 gms. (Figure 4) On cutting through multiple tissues solid grey white to grey brown friable areas with large areas of necrosis and haemorrhage was seen. Histopathologically, sections from multiple tissue pieces showed variable morphology. The tumor cells were spindled arranged in whorls and palisading pattern. (Figure 5a,b). The tumor cells were pleomorphic, spindle to serpentine in shape with hyperchromatic nuclei and light stained ill-defined cytoplasm (Figure 5 c, d). Prominent nuclear palisading was seen at the edges of geographical necrosis and at placed showed hemangiopericytoma like areas and hyalinised blood vessels. Brisk mitosis was observed. A histological diagnosis of malignant spindle cell mesenchymal tumor was made with possibilities of MPNST and Leiomyosarcoma. IHC was done for definite diagnosis which revealed tumor cells positive for S-100 (Figure 6).

DISCUSSION

Malignant peripheral nerve sheath tumor, a.k.a 'malignant schwannoma', 'neurogenic sarcoma' and 'neurofibrosarcoma', are extremely uncommon tumors which shows differentiation towards cells intrinsic to the peripheral nerve sheath. MPNSTs comprise approximately 10% of all soft tissue sarcomas. Majority of MPNSTs arise in association with major nerve trunks, including brachial and sacral plexus or sciatic nerve. Malignant peripheral nerve sheath tumours are also seen to develop secondary to prior radiation therapy of more than 10 years of latency period. There is high fold increased risk of developing MPNST in patients with von Recklinghausen disease (VRHD) when compared to the general population [4]. The mean age of presentation of MPNST in patients with VRHD is 28.7 years while it is 39.7 years in patients without association to this disease. A total of nine cases of MPNST of cervix have been reported in literature till date. The patients ranged in age from 25 to 73 years (mean 50 years).[5] No predilection for patients with either of the neurofibromatosis syndromes was evident from a review of the reported cases. Patients with VRHD and tumour size more than 5 cm and incompletely resected tumours have the poorest prognosis. Certain findings raise the suspicion of a malignant tumor as the malignant and benign lesions cannot be reliably distinguished by imaging criterias. One of the main distinguishing features is that malignant neural tumors tend to be larger than 5 cms. Ill-defined margins as another feature suggest infiltration of adjacent tissues and associated oedema. Variable features with classical central necrosis on cross-sectional imaging are common finding in malignant lesions although benign lesions with degeneration can also have a heterogeneous appearance. Calcifications are also more often seen to be associated with malignant lesions however can also be present commonly in ancient schwannomas.[6] Magnetic Resonance Imaging is the investigation of choice because it can reveal the nerve of origin and its relationship to adjacent structures [7]. Histologically, the tumor is composed of spindly to serpentine shaped cells arranged in whorles and fascicles. The spindle cells also exhibit nodular, storiform, herringbone patterns. Pseudocystic change or large areas of haemorrhage are commonly found. Stromas with alternating hypocellular areas that may be myxoid, fibrous or edematous are also commonly encountered. Varying degrees of atypical mitosis and geographical necrosis is typical [8]. Multimodality approach to management is involved in patients with MPNST. Complete resection with negative margins followed by adjuvant irradiation is associated with improved local spread of disease. Response to chemotherapy is not good [9].

There are many spindle cell tumours in differential diagnosis of MPNST commonest being leiomyosarcoma followed by fibrosarcoma, monophasic synovial sarcoma, spindle cell melanoma and undifferentiated endocervical stromal sarcoma. Immunohistochemical stains in Leiomyosarcoma shows strong positivity for Desmin and SMA. EMA and CK 7 show positivity in synovial sarcoma and melanocytic markers S-100 protein, HMB-45 and Melan-A for spindle cell melanoma and CD10 is helpful for the diagnosis of an endometrial stromal sarcoma. S-100, myelin basic protein, Leu-7 can be used to identify origin from nerve sheathdifferentiation and are immunoreactive for Vimentin as well [10]. To summarise, from our findings compared with nine previous cases, we must say that the mean age of the previously reported cases were 46.5 years and the most commonly presenting symptom was vaginal bleeding and all of them having polypoid masses. Our patient was a young 22-year-old girl and her presenting symptoms were vaginal bleeding and fever. She had the largest tumour (9.5 cm) size and an infiltrative tumor in contrast to other patients.

CONCLUSION

The diagnostic dilemma is likely to arise from the failure to recognize their nerve sheath differentiation. Subtle morphologic clues like nuclear palisading and alternating hypocellular and hypercellular areas are present clinching on the diagnosis, however which should cause one to include the appropriate markers in an immunohistochemical panel investigating an apparent cervical spindle cell sarcoma.

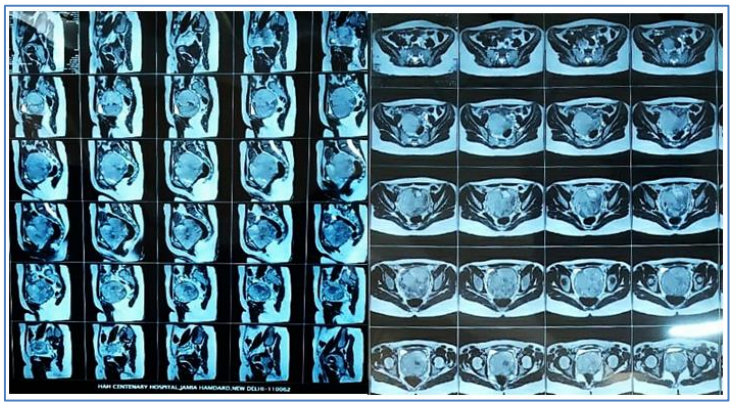

Fig-1, 2: Abdominopelvic MRI scans showing a large cervical mass exerting pressure on the bladder and rectum

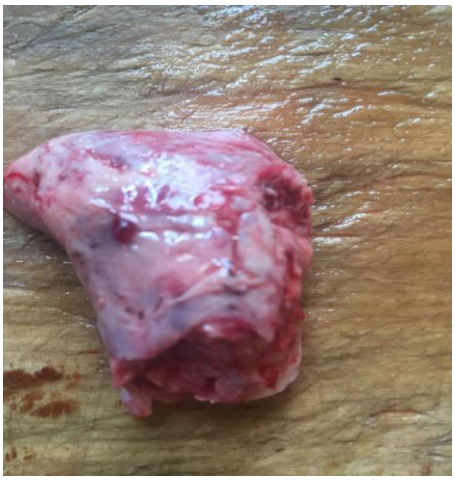

Fig-3: Fleshy polypoidal mass for performing frozen sections.

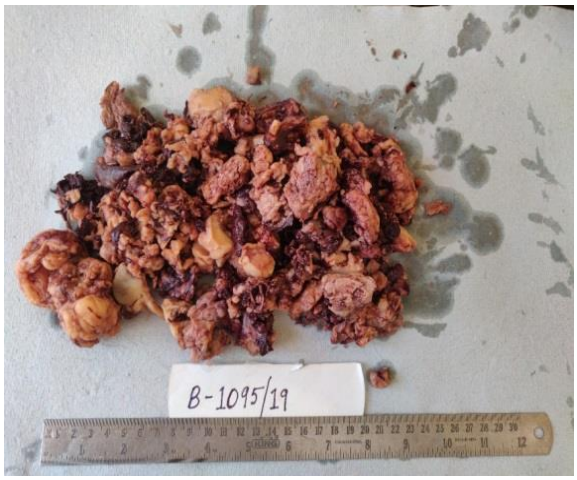

Fig-4: Grossly showing multiple irregular grey brown to grey tan soft tissue masses weighing 500 gms

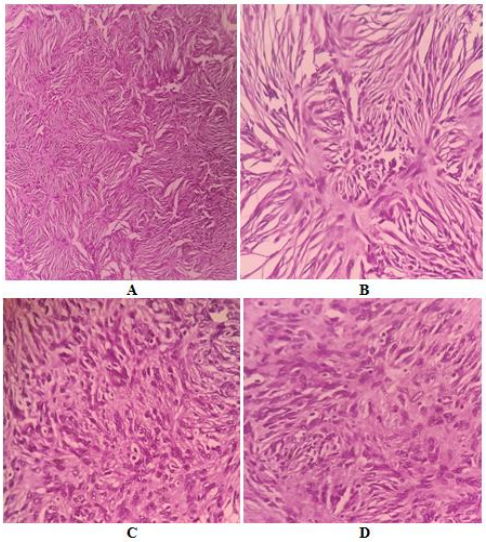

Fig-5: Photomicrograph showing tumor cells spindly arranged in whorls and palisading pattern. (a,b) The tumor cells showed moderate pleomorphism, spindle to serpentine in shape with hyperchromatic nuclei and light stained ill-defined cytoplasm. (c,d).

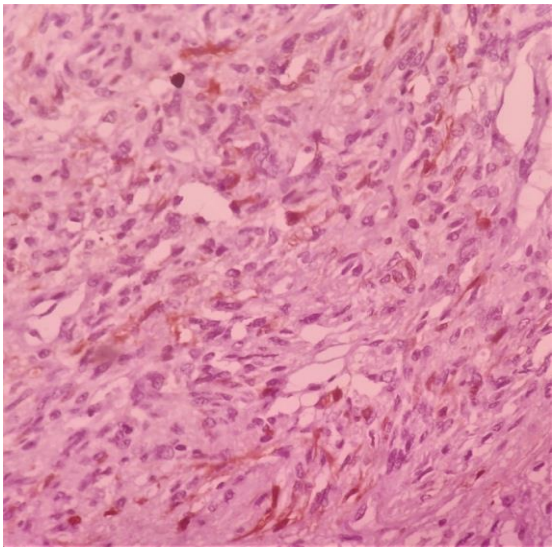

Fig-6: Photomicrograph showing tumor cells showing positivity for S-100 immunohisto Chemical stain

REFERENCES

1. Patil K, Mahima VG, Ambika L. Malignant peripheral nerve sheath tumour: An elusive

diagnosis. Indian Journal of Dental Research. 2007 Jan 1;18(1):19.

2. Kar M, Deo SS, Shukla NK, Malik A, DattaGupta S, Mohanti BK, Thulkar S. Malignant peripheral nerve sheath tumors (MPNST)–clinicopathological study and treatment outcome of twenty-four cases. World Journal of Surgical Oncology. 2006 Dec;4(1):55.
3. Ferner RE, Gutmann DH. Nerve sheath tumors in neurofibromatosis 1 international consensus statement on malignant peripheral. Cancer Res. 2002;62:1573–7.
4. Ducatman BS, Scheithauer BW, Piepgras DG, Reiman HM, Ilstrup DM. Malignant peripheral nerve sheath tumors. A clinicopathologic study of 120 cases. Cancer. 1986 May 15;57(10):2006-21.
5. Lin J, Lu C, Tsai T, Ma H. Malignant peripheral nerve sheath tumor (MPNST) involving the sacrum. Journal of medical sciences-taipei. 2007 Aug 1;27(4):179.
6. Özel E, Pestereli HE, Simsek T, Karaveli FS, Trak Bİ. Malignant peripheral nerve sheath tumor of the vulva: A case report. Turk J Cancer. 2006 Jan 1;36:31-4.
7. Lin J, Martel W. Cross-sectional imaging of peripheral nerve sheath tumors: characteristic signs on CT, MR imaging, and sonography. American Journal of Roentgenology. 2001 Jan;176(1):75-82.
8. Aisheng Dong, Changjing Zuo, Yang Wang, Zhijun Zhai and Mingjuan Xu, Enhanced CT and FDG PET/CT in Primary Malignant Peripheral Nerve Sheath Tumor of the Uterine Cervix. Clinical Nuclear Medicine. 2014; 39(9):825.
9. Rekhi B, Ingle A, Kumar R, DeSouza MA, Dikshit R, Jambhekar NA. Malignant peripheral nerve sheath tumors: Clinicopathological profile of 63 cases diagnosed at a tertiary cancer referral center in Mumbai, India. Indian Journal of Pathology and Microbiology. 2010 Oct 1;53(4):611.
10. Nielsen GP, Antonescu CR, Lothe RA. Malignant peripheral nerve sheath tumour. WHO classification of tumours of soft tissue and bone, 4th ed. Lyon: IARC. 2013:187-9.