INTRODUCTION

Majority of paraspinal tumours are benign without intraspinal invasion. The diagnosis of such lesions is usually delayed and patients remain asymptomatic with large tumour size at presentation. Epithelioid hemangioendothelioma are angiocentric vascular neoplasms with tendency to metastatize. It is one of the rarest tumours to be thought of. Its occurrence in spinal region is still exceptional. Das et al (2017) stated that only 52 cases are documented till now in English literature, with the earliest one being accounted by Maruyama et al., in 1985. The treatment modality preferred is surgical resection (gross total excision with negative margins) followed by chemoradiation.

CASE REPORT 1

A 21 year old young male patient presented with a complaint of progressively increasing swelling on the left side of the neck predominantly in the posterior region over the period of four to five months along with restriction of neck movements along with pain and numbness in left upper limb. Physical examination showed a firm mass occupying the left posterolateral aspect of the neck. On neurological examination there was no deficit. Contrast enhanced MRI of cervical spine revealed a large soft tissue intensity mass of 10x7x5 cm along the posterior paraspinal muscles on the left side closely abutting the laminae and spinous process as well as facet joint extending from C2-C7 appearing hypointense on T1 and heterogeneously hyperintense on T2W and STIR. Lesion is also encroaching on nerve exit zone causing stenosis of exiting nerve roots, with partial erosion on spinous process of C2-C3 vertebral bodies. [Figure 1] Patient was planned for gross total excision under general anaesthesia in prone position. A C-shaped incision based on the midline was placed extending from along the spinous process of C2 to C7 vertebrae. A large well encapsulated mass was present within the paraspinal muscles adherent to the lamina a spinous process of C3-C4. No evidence of encroachment into spinal canal was found. En mass excision of the tumour was done [Figure 2].  
In the early Post-operative period patient had motor loss in the left upper limb which gradually improved. After 12 days of hospitalisation patient was discharged and pathological diagnosis was followed with department of pathology. On histopathological examination the diagnosis Epithelioid Hemangioendothelioma was made [Figure 3]. Postoperative recovery was uneventful, patient received three cycles of chemotherapy. Patient was under follow up for six months when he had respiratory complaints. Contrast computed tomography (CT) chest was done which revealed multiple lung metastases as shown in [Figure 4]. Subsequently he was advised Adjuvant
radiotherapy. He is in regular follow up for last one and half year.

**CASE REPORT 2**

A 32 year old male patient presented with abdominal pain on and off for last six months with severe left lower limb pain for last four months. Upon the examination-power left lower limb 4+/5, rest 5/5. Sensory level at T12, DTR 1+ in Bilateral knee joint and ankle. Magnetic resonance imaging dorsal (MRI DL) spine shows a large well defined hypodense lesion in left hemithorax in peravertebral region. No significant arterial vascularity noted within the lesion however few small vessels from the posterior intercostals arteries are seen coursing at the periphery of the lesion [Figure 5a, b]. Patient was planned for gross excision under general anesthesia in prone position. A, T shaped incision was performed based on the midline with extension to the left side extending from along the spinous process of T10 to L1 vertebrae. A large well encapsulated mass was present within the paraspinal muscles adherent to the lamina a spinous process of T10 to L1 vertebrae. No evidence of encroachment into spinal canal was found. En mass excision of the tumour was done [Figure 6a, b]. In the early postoperative period patient had motor loss in the right lower limb which gradually improved. After seven days of hospitalisation patient was discharged and histopathological examination reveals Epithelio Hemangioendothelioma [Figure 3a, b]. Postoperative recovery was uneventful; he received three cycles of chemotherapy. Patient was under follow up for one year and no recurrence occurs.

**CASE REPORT 3**

A 28 year old female with history of pain in the bilateral lower limb (left > right) with weakness in the left lower limb for last three months. The inspection of patient revealed-power left lower limb EHL 4+/5, left knee flexion 4+/5, rest 5/5. Sensory level at L2, DTR 1+ in bilateral knee and ankle joint. Magnetic resonance (MR) defined morphology dumbell is suggestive shape of altered signal intensity mass lesion in right paravertebral region possibly arising from right neural foramen causing its widening at the level of L2-L3 showing heterogeneous post contrast enhancement and extending into the spinal canal (Intradural extramedullary) causing marked nerve root compression as shown in [Figure 7a and b]. Patient was planned for gross total excision under general anaesthesia in prone position. A, T shaped incision based on the midline with extension to the right side extending from along the spinous process of L1 to L4 vertebrae. A large well encapsulated mass was present within the paraspinal muscles adherent to the lamina a spinous process of L1 to L4 vertebrae. There is evidence of encroachment into spinal canal with intradural extension. En mass excision of the tumour was done (figure 8a, b). Postoperative period patient was moving bilateral lower limb. After 14 days of hospitalisation patient was discharged and pathological diagnosis was followed with department of pathology. On histopathological examination the diagnosis Epithelio Hemangioendothelioma was made [Figure 3a, b]. Postoperative recovery was found uneventful, patient received cycles of chemotherapy. Patient was assessed during follow up for a period of two years and no recurrence was reported.

**CASE REPORT 4**

A 26 year old female with history of upper back pain and bilateral lower limb pain from last three months. The inspection of patient revealed, power-5/5 at all joints of upper limb and lower limb power 3/5 bilateral, tone found normal, sensory was normal, reflexes were 2+ at upper limb and lower limb. A well-defined altered signal intensity lesion extending from T2-3-4-5 level causing widening and scaloping of lamina and pedicle at this levels and forming posterior mediastinal mass with compression effect on right upper lobe of lung (size – 70x77x73mm) as shown in figure 9a-9d. Patient was planned for gross total excision under general anaesthesia in prone position. A, vertical linear shaped incision based on the midline along the spinous process from C7 to T6 vertebrae. A large well encapsulated mass was present within the paraspinal muscles adherent to the lamina a spinous process of C7 to T4 vertebrae. There is evidence of encroachment into spinal canal without intradural extension. En mass excision of the tumour was done [Figure 10].

![Figure 1: (1a). MRI T1w sagittal section a paraspinal lesion extending from C3 to C5 which is isointense on T1w Imaging. Lesion is isointense with areas of hypointensities. (1b) Lesion is enhancing on contrast with areas of non-enhancement. (1c) T2w Imaging Axial Sections the lesion is having variegated appearance, with the lesion abutting the lamina and pedicles and invading the exiting nerve roots on the left side. (1d) there is no intraspinal extension however the bone and the peristomeum are enhancing.](image-url)
Figure 2: Removed en mass from left paraspinal region (11x8 cm)

Figure 3: (a) microscopically, tumour was composed of small nests of rounded to slightly spindled endothelial cells embedded in a light basophilic myxoid matrix. Cells had a round regular nucleus and vacuolated cytoplasm. (b) The Lesion was CD34 positive which is seen in almost 80 percent of the cases.

Figure 4: High-resolution computed tomography (HRCT) lungs revealed multiple pulmonary metastasis

Figure 5: (a, b) Magnetic resonance imaging of dorsal spine (MRI DL spine)

Figure 6: (a) Removed en mass from left paraspinal region. (b) Postoperative recovery of patient

Figure 7: (a, b) Magnetic resonance imaging of Lumbosacral Spine (MRI LS Spine)

Figure 8: (a, b) Removed en mass from right paravertebral region

Figure 9: (a-d) Magnetic resonance imaging of cervicothoracic region
Postoperative period patient was moving bilateral lower limb. After 14 days of hospitalisation patient was discharged and pathological diagnosis was followed with department of pathology. On histopathological examination the diagnosis Epithelioid Hemangioendothelioma was made. Postoperative recovery was found uneventful, patient received cycles of chemotherapy. Patient was assessed during follow up for a period of six months and no recurrence was reported.

DISCUSSION

A paraspinal mass may be caused by large number of conditions including an abscess, benign and malignant neoplasm involving mesenchymal tissue. The diagnosis of many such lesions may be done with ease especially when correlated with clinical and radiological findings. Paraspinal tumours can develop and grow into a more abundant space than intraspinal tumours. Also, these tumours are slow growing and attain a large size at the time of diagnosis. We hereby report a rare case of paraspinal mass which is diagnosed clinically as liposarcoma or myosarcoma or neurofibroma but the histopathological diagnosis was Epithelioid hemangioendothelioma.

Epithelioid hemangioendothelioma are well differentiated endothelial soft tissue tumours. Mostly affects lung, liver and bones but may also involve head and neck areas, mediastinum, lymph nodes, soft tissue and spine. Its occurrence in spinal and paraspinal areas is least documented. According to WHO classification (2020), of tumors of soft tissue and bone, epithelioid hemangioendothelioma are malignant vascular tumours composed of epithelioid endothelial cells with distinct myxohyaline stroma and they are arranged in short cords. EHEs mostly occur in young aged individuals with no gender predilection. Prevalence is less than 1/10, 00,000. Patients present with progressive pain and if they arise in vertebrae they might cause spine compression and therefore paraesthesia, loss of muscle strength and paraplegia. EHE can be unifocal, locoregional spread or systemic involvement. A radiographic finding of the EHE is variable. MRI is imaging modality of choice for diagnosis, the relations with the surrounding tissue and a potential cleavage plan. Mostly EHE are hypointense on T1-weighted (T1W) and hyperintense on T2W and homogenous enhancement after contrast enhancement, as > 50 % of the cases have systemic involvement at presentation fluorodeoxyglucose-positron emission tomography (FDG-PET) computed tomography(CT) is advisable with mild to moderate uptake.

In pathological studies, the tumour may be well circumscribed or indistinct borders varying from light purple to red, soft to firm in consistency. Microscopic analysis shows cells with round to oval nucleus with intracytoplasmic vacuolations. The tumour is characterized by positive endothelial cell markers such as CD31, CD34 and factor VIII related antigen which are positive in less than 30% cases.

Nuclear expression usually shows positivity for calmodulin binding transcription activator 1 (CAMTA1), WW domain containing transcription regulator 1 (WWTR1), and TFE3. Tumour is graded on the basis of combination of mitotic activity and tumour size or histological atypia and tumour size. In view of the fact that of its heterogeneous presentation and accountability of less than 1% of all vascular tumours, it is misdiagnosed and not suitably treated leading to poor prognosis. Owing to the rare occurrence of the tumour, there is limited documentation regarding standard treatment modality for these tumours. Gross total resection with negative margins remains the standard treatment modality. Adjuvant chemoradiotherapy is also known as adjuvant therapy.

In our case, the patient was managed by gross total resection of tumour with negative margins with no spinal stability intervention followed by adjuvant chemotherapy and external beam radiotherapy. Patient is clinically stable and on regular follow up. There can be a number of complications occur during resection of paraspinal tumours according to tumour location, which include laryngeal paralysis, neurogenic bladder, femoral nerve palsy and rectal dysfunction. In our case, motor weakness is the only surgical complication.

CONCLUSION

We believe that Epithelioid hemangioblasto is infrequent in the paraspinal area. It is difficult to make a diagnosis before surgery due to lack of characteristics clinical picture and radiological findings. En block resection of tumour with negative margins can possibly be practised. The combined therapy of chemo radiation and surgery is yet to studied upon, as any period of follow up is too short.
to determine the tumour recurrence, it needs a prolonged follow up and surveillance. Although rare, EHE should be included in differential diagnosis of paraspinal tumours. Spinal EHE, though a rare entity, is becoming familiar with more and more cases being reported. The best possible control methods are yet to be suggested depending on clinical assessments. In the current scenario, surgical resection, oncologic margin clearance as well as functional preservation is the target, with radiation being retained for cases with an incomplete removal or with tumours at unreachable locations. Preoperative angiogram and embolization must be implemented, if possible.

REFERENCES