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Case Report

Presacral Hemangiopericytoma: Case Report

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Abstract

Hemangiopericytoma is rare tumor originating from the pericytes, the contractile cells that surround capillaries. This tumor arises from pericapillary cells or pericytes of Zimmerman and can occur anywhere where capillaries are found. However hemangiopericytoma develops mainly in the lower extremities, retroperitoneum or pelvis. The recommended treatment for a hemangiopericytoma is wide excision. Since many benign and malignant diseases occur in the area surrounding the anus, recognizing the presence of this malignant tumor in the presacral space is important in order to avoid inappropriate surgery such as piecemeal excision. We describe a rare case of a hemangiopericytoma in the presacral space. With posterior approach the tumour was completely removed. Pathological examination including immunohistological stains was consistent with a hemangiopericytoma.

Keywords: Hemangiopericytoma, Presacral, Surgery, Immunohistology.

1. Introduction

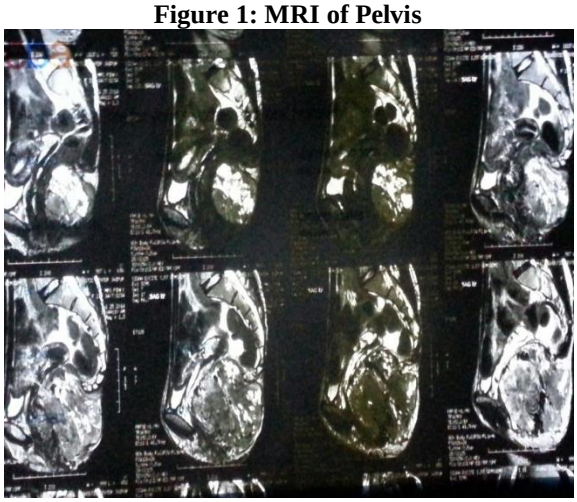
Presacral tumors are rarely encountered in surgical practice. A presacral hemangiopericytoma is a rare, soft tissue tumor of vascular origin derived from a pericyte of Zimmerman, which is a modified smooth muscle cell that surrounds the small blood vessels. This type of tumor was first described by Stout and Murray in 1942[1]. It represents approximately 5% of all sarcomatoustumors and can occur anywhere but more usually in the musculature of the extremities, retroperitoneum, pelvis, head, neck and lungs [2]. The rarity of its occurrence and its anatomical site may make it difficult for the examining surgeon to diagnose and treat this disorder. Of a variety of surgical approaches advocated for presacral tumors, a growing number of posterior approaches have recently been selected[3]. Complete excision has a favourable effect on recurrence and survival. It improves survival and reduces the chance of first recurrence. Since the recommended treatment for a hemangiopericytoma is wide excision, due to high local recurrence it is important to recognize the presence of this malignant tumor in the area surrounding the anus, where various tumorsoccur[4]. Histopathology reveals spindle-shaped cells with multiple vascular spaces. Hemangiopericytomas are classified as benign or malignant. Malignant hemangiopericytoma is characterized by the presence of necrosis and or more than five mitoses per ten 400 microscopic fields and at least two of the following microscopic features: haemorrhage, moderate to-high nuclear atypia, or moderate-to-high cellularity. Immunohistochemistry is positive for vimentin, S-100, smooth muscle actin, CD34. Presence of mitoses, necrosis, and vascular invasion predicts aggressive behavior and poor prognosis.

2. Case Report

A 25 years old female presented in outpatient department of general surgery at SMS Hospital with chief complains of swelling over sacral area for 4 month and pain in swelling for 2 month. There was no history of fever, bleeding per rectum, constipation, urinary problems, pain or weakness in lower limbs, weight loss. On local examination swelling present over the sacral area 9×9 cm in size, oval in shape, smooth surface, firm in consistency, mobile and not fix to the skin. Patient was admitted

in hospital and all routine blood investigation was within normal limit including carcinoembryonic antigen. On rectal digital examination, elastic hard, painless mass was palpate right posterior wall of the rectum. Proctoscopy revealed a bulging of the posterior rectal wall toward the luminal face. The mucosa was normal. Endorectal ultrasound showed a 9×8 cm solid lesion with external compression of the posterior wall of thelower rectum, with no signs of invasion of the rectal wall. Magnetic resonance imaging showed a Large heterogeneous lobulated predominantly solid having small cystic/necrotic areas measuring about $10 \times 10 \times 10$ cm in perineum and lower pelvis region in bilateral ischio-rectal fossa and right perineal region in posterior and lateral aspects of lower rectum and anal canal causing anterior displacement of anal canal, lower rectum, vagina and right levetorani muscle likely suggestive of soft tissue tumor (figure 1). Multiple vascular flow voids are seen within and around the mass.Pre-operative biopsy was not advocated. The patient underwent surgery and the mass was easilydissected via a posterior sagittal approach. Intraoperatively, the solid mass 10×10 cm appeared to be an elastic tissue that was well encapsulated present in presacral area pushing the rectum and anal canal anteriorly with extensive vascularity. There was noevidenceof invasion of the rectum, sacrum, or coccyx.The patient made an uneventful recoveryand was discharged on the 6th postoperative day with stable vitals.

Figure 1: MRI of Pelvis



Large heterogeneous lobulated predominantly solid having small cystic/necrotic areas measuring about $10 \times 10 \times 10$ cm in perineum and lower pelvis region, suggestive of soft tissue tumor.

Histopathological assessment of the specimen revealed a10×9×9 cm mass with a thin capsule. On sectioning, the cut surface of the mass was yellow brown with numerous dilated vascular spaces. Microscopically the tumour consisted of tightly packed, round to fusiform cells with indistinct cytoplasmic borders that were arranged around an elaborate vasculature (figure 2). On follow up tumor marker study showed following finding (table 1). Patient was referred to medical oncology department for further management.

Figure 2: Histopathology showing multiple vessels surrounded by spindle cells.

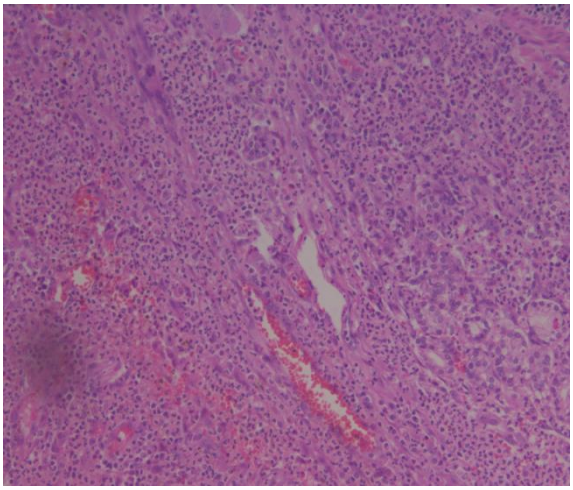


Table 1: Immunohistochemistry (IHC) report

IHC Marker	Result
Vimentin	Positive
CD 34	Positive
Smooth Muscle Actin	Negative

3. Discussion

Hemangiopericytoma is an indolent slow growing tumor in its classical presentation with a high recurrence rate. As originated in the capillary pericytes it can arise in any part of the body.Clinicalpresentation is non-specific. Pain is a late symptom associated with an enlarging mass. Most tumours present asa slowly growing mass that may cause intestinal orurinary symptoms in presacral region. Rectal examination is the mostimportant most effective and least expensive means of identifying presacral tumours. Colonoscopy can rule out any rectal mucosal changes in cases of rectal bleeding. Endorectal ultrasound (ERUS) has been used to characterize the lesion as solid or cystic and it occasionally shows internal echoes due to mucoid or inflammatory debris. Computed tomography (CT) and magnetic resonance imaging (MRI) are the most accurate diagnostic tools. Presacral tumors generally appear as well-circumscribed, radiopaque soft tissue mass lesions that often displace adjacent structures.

However, radiologic diagnosis with CT or MRIis rarely in agreement with the final histology. Pre-operative definitive diagnosis of presacral masses

with non-invasive study is difficult. Generally preoperative biopsy is not preferred, due to the risk of spreading dysplastic cells. Furthermore, the tissue obtained is often not enough to make a definitive histological diagnosis. Although in some cases pre-operative biopsy is usually recommended for unresectable lesions for neo-adjuvant treatment. Complete excision is necessary to prevent recurrence, infection, and possible malignant transformation. If the tumor does not extend above the level of the 4^{th}sacral element, a posterior approach is the appropriate method. An anterior or abdominal approach is preferred for lesions with the lowest extent above the 4th level of the sacral element. If the lesion begins under the 4^{th}level of the sacral element and extends well above it, a combination approach may be necessary[5]. The posteriorsagittal approach offers the advantages of avoiding the risk of intra-abdominal complications, reducing patient discomfort, and good surgical exposure.

In our case the tumor was excised completely and easily by a posterior approach, without any complications. Cases of hemangiopericytoma undergoing complete tumor resection achieved a 100% survival rate at five years. Hemangiopericytoma is capable of both local recurrence and distant metastases, but has low disease associated mortality[6]. The most common metastatic sites by the haematogenous route are the lungs, liver and bone. Local and distant relapses after a prolonged disease free interval have been reported suggesting mandatory long term follow-up. Adjuvant chemotherapy and radiotherapy are recommended inpatients with incomplete resection and or large locally invasive tumours.

Preoperative radiotherapy is indicated in high-risk surgical cases and surgically inaccessible tumors. Patients reported that in patients who received preoperative radiotherapy, removal of tumor was easier with less hemorrhage. Histological examination post radiation showed reduction in tumor cells with hyaline degeneration[7]. Postoperative radiotherapy is associated with better survival and have recommended prophylactic postoperative radiotherapy after complete tumor excision to reduce local recurrence rates. A dose of 50 Gy is recommended after first tumor excision. The role of chemotherapy is more in the palliative setting than as primary treatment. Treatment regimens commonly include actinomycin D, adriamycin, vincristine, and cyclophosphamide and methotrexate. Response rates to chemotherapy range between 30-40% and less than 50% are disease free at 5 years after diagnosis of metastasis[8]. Although the patient is tumourfree at 6 month follow-up further follow-up is mandatory.

4. Conclusion

This report presented a rare case of a hemangiopericytoma in the presacral space. Many diseases are associated with anal lesions; therefore a thorough differential diagnosis and complete resection without piecemeal excision must always be performed in the management of this type of malignant tumor.

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