CASE REPORT

Hemangiopericytoma in Perineal Region

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Introduction

Hemangiopericytoma is a rare soft tissue tumor of vascular origin which is derived from pericytes. The pericyte is a smooth muscle cell that surrounds small blood vessels. This tumor was first described by Stout and Muary in 1942. It is usually seen in the musculature of extremities, the retroperitoneum, pelvis, head, neck, and lungs. There is only one report of hemangiopericytoma in the sacral space [1]. The recommended treatment for a hemangiopericytoma is wide excision due to high local recurrence. Two-third of these tumors are benign, while one-third are malignant. We hereby report a rare case of hemangiopericytoma in the perineal space.

Case Presentation

A 34-year-old female patient, P₁A₀L₁, got admitted at the Government Medical College, Haldwani, with complaints of a large mass in the left vaginal and pelvic area of about 8–10 cm, which was gradually increasing in size for the past 3–4 months, on and off fever for the past 1 week, difficulty in walking and carrying out routine activities for the past

3–4 days, and acute retention of urine for the past 2 days. There was no significant past medical or surgical history. On admission, her general condition was average, febrile, dehydrated, no pallor, no icterus, no edema, and no lymphadenopathy. Her details were BP: 136/90 mmHg; PR: 90/min; and RR: 17/min. On per abdominal examination, tenderness was present and the lump palpable in the suprapubic area (?Distended bladder), with no guarding, no rigidity. On per speculum examination, the cervix and vagina were healthy, with dirty discharge (++). On per vaginal examination, a large firm mass was felt in the left side of the vagina extending into the perineal region $\approx 10 \times 10$ cm, with no obvious tenderness; the uterus size could not be made out.

All routine investigations were carried out including blood tests—hemogram, liver function tests, kidney function tests, coagulation profile, ECG, and chest X-ray. Ultrasonography of the whole abdomen including the uterus, ovaries, kidneys, and urinary bladder was normal. TVS showed evidence of a highly vascular tumor between the posterior layer of the bladder and the anterior layer of the rectum along with effacement of the left ischiorectal fossa. The tumor was well encapsulated and the rectum was intact. MRI showed that the outer layer was heterogenous with high intensity and that the central layer had an extremely high intensity in T2 weighted images, indicating that the inside of the mass had a rich blood flow. Biopsy was not taken as it was associated with needle track seeding.

Because of its vascularity, pre-operative diagnosis of Rhabdomyosarcoma, solitary fibrous tumor, fibrous histiocytoma, and hemangiosarcoma was made. After all the

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hemangiopericytoma(gross)

Fig. 1 Hemangiopericytoma (gross) **Fig. 2** Hemangiopericytoma (microscopic-H&E stain)

pre-operative workup, the patient was taken for a wide local excision of the tumor. The patient underwent a tumorectomy via perineal incision. The tumor was completely removed and sent for histopathology. Its cut surface was grayish white.

Perop—during the procedure, the patient started bleeding profusely and 2 units of whole blood were transfused. The patient went into cardiac arrest on the operating table; she was adequately resuscitated and after insuring complete hemostasis, she was shifted to the critical care ward. However, her condition started deteriorating and her vitals started falling. Bleeding continued from the perineal region. The patient was again shifted to the operation theater after about 2 h, the perineal stitches were removed, and the bleeders were caught and ligated. Bleeding still continued and a laparotomy was planned. Bilateral internal iliac arteries' ligation was done. Complete hemostasis was achieved and tight vaginal packing was done. The patient was intubated and shifted to the ICU where she was put on ventilatory support. Unfortunately, she continued to bleed and started going into shock and was again shifted for exploration to the operation theater after 4 h. Again, the perineum was re-explored, all the bleeders were caught and ligated successfully, and she was shifted to the ICU on the ventilator. During this whole period, she was transfused another 6 units of whole blood. She was transfused 3 units of whole blood and 8 units of fresh frozen plasma in the post-operative period in the ICU. Her condition continued to improve; she was taken off the ventilatory support after 3 days and was shifted to the general ward after 4 days of ICU care. She was discharged in a satisfactory condition after complete stitch removal and adequate advice.

Discussion

Very little has been published about hemangiopericytoma.

It is a rare soft tissue tumor of vascular origin which is derived from the pericytes and can occur in any area where vascular capillaries are found. Since their original description in the female genital tract in 1954, a majority of these lesions have been of uterine origin. It usually presents as a painless growing mass; however, pain may result from compression of adjacent nerves. There have been great problems in formulating reliable criteria for differentiating benign from malignant hemangiopericytoma [2].

We diagnosed hemangiopericytoma following an examination of structural features of the mass. Spindle-shaped cells surrounding endothelial lined vascular spaces were observed by H&E staining. Histopathologic examination revealed that the tumor contained spindle-shaped cells surrounding the endothelial lined vascular spaces, which is consistent with histology with hemangiopericytoma. The mass was positive for CD 34 and vimentin.

Radiotherapy and chemotherapy are generally not effective for treatment of hemangiopericytoma. Tumor is radioresistant and out of six patients with pre-operative chemotherapy, only one responded to anthracycline-based

chemotherapy. The 5-year survival rate in patients treated with curative resection and non-curative resection was 79 and 50 %, respectively. All resectable hemangiopericytoma should be treated with wide local excision. Radiotherapy and chemotherapy have been traditionally reserved for non-resectable and metastatic cases. Hemangiopericytoma is a diagnosis of exclusion and surgical resection is the only way of achieving a cure [3]. These data indicate that a complete resection is necessary to improve patients' survival rates as was done in our case. This case report presented a rare case of hemangiopericytoma in the perineal region (Figs. 1, 2).

References

1. Kitahata Y, Yokoyama S, Takifuji K, et al. Hemangiopericytoma in the sacrococcygeal space: a case report. *J Med Case Rep.* 2010;4:8.
2. Singh V, Raghavendran M, Kapoor R. Hemangiopericytoma of pelvis: a case report. *Indian J Urol.* 2003;20:62–3.
3. Goldman SM, Davidson AJ, Neal J. Retroperitoneal and pelvic hemangiopericytoma: clinical, radiologic, and pathologic correlation. *Radiology.* 1998;168:13–7.