Management of presacral tumors: Our experience with posterior approach

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1. Introduction

Presacral tumors arise in the presacral or retrorectal space. The boundaries are comprised by the fascia propria of the upper 2/3rd of rectum anteriorly, presacral fascia posteriorly and the endopelvic fascia with the ureters and iliac vessels on both lateral aspects. The roof is formed by the peritoneal reflection while the floor by the Waldeyer’s fascia. It constitutes an area for development of multiple embryologic structures between the hindgut and the proc-todeum, neural elements and bone and connective tissue. Many tumors, mostly congenital in origin, may arise from these tissues. The incidence has been reported in literature to be between 1 in 40,000 to 1 in 63,000 hospital admissions [1,2]. Due to obscure anatomic location, difficult surgical approach and etiological heterogeneity, tumors arising here pose a diagnostic and therapeutic challenge. We present our experience with 10 cases of presacral tumors with posterior approach being the major surgical approach.

2. Materials and methods

All patients of presacral tumors presenting in the Department of General Surgery, Sawai Man Singh Medical College in a single surgical unit during a time period of 14 months (May 2013–July 2014) were included in the study. Data was collected based on the complete clinical records maintained at our hospital. The clinical presentation, diagnoses, surgical intervention done, post-operative outcome, and histopathological findings were reviewed. Follow up was performed on outpatient visits.

3. Results

Total of 10 patients were included in the study. All patients were routine outpatient admissions. All the patients were females in the age group of 18–50 (mean 28.4) years. The pathological findings included schwannoma, leiomyosarcoma, hemangiopericytoma, neurofibroma, paraganglioma and rest were developmental cysts. 6 cases were managed using the posterior approach and rest by anterior approach. There was no major complication or mortality in the follow up. Complete surgical excision remains the mainstay of therapy. Surgical approach depends upon the location, size, local invasion and surgical expertise of the surgeon. Benign tumors have a good prognosis while the prognosis in malignant tumors remains guarded due to difficulty in obtaining safe resection margins. Posterior approach is an attractive option for low lying, benign tumors that is more direct, with better exposure and quicker recovery.
The most common presenting symptom was lump lower abdomen (5/10) followed by swelling in lower back and perineal region (Fig. 1). One patient presented with features of adrenergic overactivity manifested as body ache, headache and palpitations. Pressure symptoms due to mass effect were present in two patients. One had edema of the left lower limb and second had neuropathic pain in left lower limb. One patient also complained of burning micturition. Duration of symptoms ranged from 1 to 36 months (median of 3 months).

Three out of the 10 patients had been operated previously for lump abdomen and perineal swelling. Duration of symptoms ranged from 1 to 3 months. In follow up, there was no evidence of bowel or bladder dysfunction in any of the patients operated by posterior approach; neither was there any residual limb weakness or sensation loss.

The HPE findings have been illustrated in Table 2. Margins were negative.

4. Discussion

Presacral tumors are a rare variety of space occupying lesions whose true incidence is not known because most of the series have been published from tertiary care referral centers. Despite this, various authors report an incidence between 1.4 and 6.3 cases per year [2–6]. The incidence of benign tumors has been reported to be more in females while malignant tumors have a slight male predominance [2,3,4,7–9]. Several classification systems have been proposed but most commonly the retrorectal masses are classified based on their origin into congenital (55–65%), neurogenic (10–12%), osseous (5–11%), inflammatory (5%) and miscellaneous (12–16%).

The congenital tumors are most common and arise from all three germ layers. These are more common in females. These may arise from ectoderm (dermoids and epidermoids), sequestration of hindgut remnants (tail gut cysts, rectal duplication cysts), neural tube defects (anterior sacral meningocele), and notochord (chordomas). Clinical presentation is nonspecific depending on the size and location of the mass and tissue of origin. A careful digital rectal examination (DRE hereafter) is diagnostic in 90%. Chordoma is the most common malignant presacral mass.

Neurogenic lesions typically arise from the peripheral nerves. Ependymomas are the most common histological type. Neuropathic pain and nerve weakness are the presenting symptoms. They may give rise to pelvic mass effect when large in size.

Osseous tumors may be both benign and malignant. Symptoms most often arise due to compression of nearby neurological structures causing radicular pain. Biological behavior is similar to osseous tumors elsewhere in the body. Complete resection is recommended due to high rates of recurrence.

Symptoms arising due to presacral masses depend on the size, location and origin of the tumor and also on whether the tumor is benign or malignant. In a review of five largest case series, 43% of benign tumors were free from symptoms while only 7% of malignant tumors were symptomless [8,10]. A careful DRE is the initial examination of choice. In a series of 120 cases from

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**Table 1**

Demographic and clinical profile of the patients.

| S. no. | Age | Sex | Presenting complaints | Duration of symptoms (Mo) |
|--------|-----|-----|------------------------|--------------------------|
| 1      | 18  | F   | Lump abdomen, pain abdomen | 2                        |
| 2      | 45  | F   | Lump abdomen, lower limb edema | 1                        |
| 3      | 21  | F   | Lump abdomen, lower limb pain | 24                       |
| 4      | 42  | F   | Lump abdomen | 15                      |
| 5      | 18  | F   | Body ache, headache, palpitations | 36                       |
| 6      | 25  | F   | Perineal Swelling | 3                        |
| 7      | 38  | F   | Swelling lower back, pain in swelling | 5                        |
| 8      | 27  | F   | Swelling coccygeal region | 2                        |
| 9      | 30  | F   | Swelling lower back | 3                        |
| 10     | 20  | F   | Lump abdomen, pain abdomen | 2                        |

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**Fig. 1.** Preoperative photograph of the patient 7 showing swelling over lower back.
Mayo clinic, 97% of the tumors were palpable per rectally [2,10]. If the superior limit of the tumor is palpable on DRE, a posterior surgical approach may be feasible. Plain radiographs may sometimes be useful and demonstrate sacral destruction (chordomas), calcifications within the mass (teratomas) or pathognomic signs like the “Scimitar sign” (anterior sacral meningocele). Computed tomography of the pelvis is widely used and is capable of differentiating solid from cystic lesions, vascular, adjacent organ involvement and bony destruction. Contrast enhanced MRI pelvis is the gold standard investigation of choice and the most sensitive and specific investigation also. Limitations of MRI are inability to distinguish between fibrosis and recurrent pelvic carcinoma in patients managed with surgery or radiation therapy. Other investigations like CT/MR myelography may be done when there is involvement of CNS. In cases of major vessel involvement, preoperative angiography may be necessary to expose the major feeders of the tumor. Endorectal ultrasound and lower GI endoscopy can help in evaluating the involvement of layers of rectum and superior extent of the lesion as well as the consistency of the tumor [11].

The role of preoperative biopsy is controversial. A preoperative biopsy should only be performed if it is likely to change the management and surgical approach. Currently, there is no indication for biopsy of a cystic lesion [11,12]. Adequate resection is diagnostic as well as therapeutic. Preoperative percutaneous biopsy is, however, indicated for solid or heterogeneous tumors to facilitate decision making for neoadjuvant therapy and surgical planning [12]. Biopsy, if indicated, should never be done transperitoneally, transretroperitoneally, transrectally or transvaginally [11].

Three main surgical approaches are used: anterior, posterior and combined. The anterior or trans abdominal approach is recommended for very large tumors, when there is suspicion of malignancy and when the lower limit of the tumor is above the S3 vertebra. Advantages of an anterior approach include better visualization of pelvic structures, bleeding control and easy mobilization of rectum.

The posterior approach, first described by Paul Kraske (1885), may be inter-sphincteric, trans-sphincteric parasacrococcygeal, trans-sacral, trans-sacrococcygeal, trans-anorectal, trans-vaginal.

Table 2
Findings on DRE, final histopathological diagnosis, post – operative recovery of the patients.

| S. no. | Mass felt on DRE | Surgical approach | Histopathology     | ICU requirement | Duration of hospital stay (Days) | Follow up (Mo) | Bladder/bowel function | Residual nerve weakness |
|--------|------------------|-------------------|--------------------|----------------|---------------------------------|----------------|------------------------|------------------------|
| 1      | Yes              | Post.             | Schwannoma         | Yes            | 10                              | 2              | Normal                 | None                   |
| 2      | No               | Ant.              | Lymphoma           | No             | 8                               | 12             | Normal                 | None                   |
| 3      | No               | Ant.              | Neurofibroma       | No             | 7                               | 12             | Normal                 | None                   |
| 4      | No               | N/A               | Leiomyosarcoma     | N/A            | 3                               | N/A            | N/A                    | N/A                    |
| 5      | No               | Ant.              | Paragangioma       | Yes            | 9                               | 3              | Normal                 | None                   |
| 6      | Yes              | Post.             | Epidermoid         | No             | 2                               | 3              | Normal                 | None                   |
| 7      | Yes              | Post.             | Epidermoid         | No             | 2                               | 2              | Normal                 | None                   |
| 8      | Yes              | Post.             | Dermoid            | No             | 2                               | 2              | Normal                 | None                   |
| 9      | Yes              | Post.             | Epidermoid         | No             | 2                               | 2              | Normal                 | None                   |
| 10     | Yes              | Post.             | Hemangiopericytoma | No             | 7                               | 1              | Normal                 | None                   |

* Recurrent cases.
It is indicated in low lying tumors with upper limit below S3, when there is evidence of sacral involvement, in small tumors and when malignancy is not suspected. Coccygectomy – indicated for better exposure as well as for complete removal of a potential communication route and consequent recurrence of cystic lesions or teratomas. Advantages include a more direct approach, more commodious to adopt, operating field of the tumor is clearer, adverse influences of bowel coming in between are avoided, quicker postoperative recovery. Potential drawbacks include absence of control over pelvic vessels, chances of injury to pelvic nerves and reduced working space.

A combined abdomino-sacral approach is recommended for larger lesions that extend both above and below S3, large masses, and suspected malignancy.

A perineal endoscopic approach has also been described to access the presacral space and perform excision biopsies.[13]

Complications of surgery include bleeding, post-operative urinary and fecal incontinence, nerve root injury leading to weakness and chances of abscess or fistula formation. Rates of complications are higher in patients undergoing some form of sacral resection. Even, if sacrectomy is necessary, care must be taken to preserve at least one side of S2 to reduce postoperative complications arising out of nerve damage.

5. Conclusion

Adequate surgical resection is the only treatment for presacral masses as most of the tumors eventually enlarge in size; there is always a risk of malignancy and infection; and they have the potential of causing dystocia in women of child bearing age. Posterior surgical approach is an attractive option as most common variety of presacral masses is low lying developmental cysts. Rate of complications can be avoided by careful selection of cases. The prognosis of benign tumors is excellent after complete excision while that of malignant tumors is poor due to high rates of recurrence.

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Consent

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Author contribution

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Guarantor

Dr. Dhananjay Saxena, the contributing author, is the Guarantor.

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