Aims: The study aimed to evaluate the presentation, management, and outcome of patients of sacrococcygeal malignant germ cell tumors (SC-MGCTs) with intraspinal extension.

Materials and Methods: Case records of all cases of SC-MGCT were reviewed to identify cases with intraspinal extension. They were evaluated in terms of their presentation, response to therapy, extent of surgical resection, recovery of neurological symptoms if any, and outcome.

Results: Of the three cases of SC-MGCT, two had intraspinal extension. One had Altman Type 3 disease and another had Altman Type 4 disease; both had Stage 4 disease. The intraspinal extension in both patients was detected on contrast computed tomography scan and magnetic resonance imaging. One patient had bladder and bowel incontinence. All the tumors responded to preoperative chemotherapy. Gross complete local resection could be achieved in all patients. Neurological recovery was complete. All were alive with no recurrence on follow-up.

Conclusions: Malignant sacrococcygeal GCTs with intraspinal extension are rare and can be managed with neoadjuvant chemotherapy which obviates the need for extensive exploration for excision and has got good neurological and oncological prognosis.

Keywords: Intraspinal extension, malignant germ cell tumor, sacrococcygeal tumors
tomography (CT) scan performed at presentation which showed tumor extension into the sacral canal through vertebral foramina or vertebral erosion. Magnetic resonance imaging (MRI) spine was done to delineate the exact intraspinal extent of the tumor. Altman grouping (4) and Pediatric Oncology Group staging systems were used. (5) Both children received chemotherapy as per the JEB regimen – carboplatin 600 mg/m²/day on day 2 of course for 6 courses, etoposide 100 mg/m²/day for 3 days for 6 courses, and bleomycin 15 mg/m²/day on day 3 of course for 6 courses. (6) Three of these 6 courses were given as neoadjuvant chemotherapy preoperatively. Surgical excision was then performed and patients received further 3 courses of adjuvant chemotherapy of the same regimen. Surgical resection was performed through a standard inverted V incision, with excision of the residual sacrococcygeal mass and the coccyx. Patients were followed up with serum AFP and CT scans or ultrasound examinations 3 monthly for the 1st year and then 6 monthly as per the protocol.

RESULTS
Out of three children of SC-MGCT who presented to us over a span of 12 months, two children had an intraspinal extension of the tumor. The first case was a 14-month-old female child with swelling in the sacrococcygeal region. The child also had constipation and straining at micturition. Physical examination revealed a 10 cm × 7 cm swelling in the right gluteal inferomedial aspect reaching midline [Figure 1a]. Per-rectal examination revealed mass pushing into the lumen from the right posterolateral aspect with intact rectal wall and mass was having variable consistency. The AFP levels were elevated (3612 ng/ml) and beta-human chorionic gonadotropin (HCG) was normal. The second case was a 3-year-old female child who presented with constipation, urine retention, and lower-limb weakness (power 3/5). On examination, there was no external lump, but on per-rectal examination, mass of variable consistency with cystic areas felt in the right lateral aspect of the rectum with intact mucosa. Her AFP was raised (1210 ng/ml).

Younger child presented with Altman Type 3 while the older child had Altman Type 4 disease. MRI was showing sacrococcygeal mass with intraspinal extension in both children [Figures 1b and 2a]. Both were having Stage 4 disease with lung metastases [Figures 1c and 2b]. Both patients received three cycles of neoadjuvant chemotherapy [Table 1]. Both the tumors responded well to neoadjuvant chemotherapy, with the pulmonary metastasis resolving completely and primary tumor shrinking in size and the intraspinal tumor resolving completely [Figures 1e and 2c]. Complete surgical resection of the residual sacrococcygeal mass with coccyx was possible in both children without requiring any laminectomy or intraspinal dissection [Figures 1d and 2d]. Following resection, both children received three additional cycles of adjuvant chemotherapy [Table 1]. Pathology revealed sacrococcygeal germ cell tumor without any residual immature or endodermal sinus component in both children. The child with lower-limb paresis regained normal power of lower limbs, and the child with urinary incontinence showed resolution of urinary incontinence. Both children are under follow-up (12 months and 18 months, respectively) and with no recurrence.

DISCUSSION
Intraspinal extension of the isolated Sacrococcygeal

Figure 1: Case 1 (a) Clinical picture of sacrococcygeal mass, (b) magnetic resonance imaging showing sacrococcygeal mass with intraspinal extension (arrow), (c) chest computed tomography showing bilateral lung metastases (arrows), (d) excised tumor along with coccyectomy, (e) resolved intraspinal extension post chemotherapy
Sacrococcygeal teratoma (SCTs) is relatively rare. The majority of the reported cases of SCT with intraspinal extension have been mature or immature teratomas. These cases did not present with neurological deficits and were excised in the neonatal period in toto including the intraspinal extension. Seifern Und Aspang et al. reported three neonatal SCTs with intraspinal extension; two of them had raised AFP than normal for age. Although all three were excised in toto in the neonatal period, one required sacrifice of sacral nerve root and developed paraparesis and neurogenic bladder and one neonate required laminectomy for complete excision and subsequent neurological deficits occurred.

Our cases are unique in that both were malignant GCTs presenting late beyond infancy with neurological symptoms in the form of bladder incontinence or lower-limb paresis.

Sacrococcygeal GCTs present with constipation and/or urinary retention, paraparesis, and dribbling of urine. One should not miss per-rectal examination. The child who presented at 3-year of age was in fact an Altman Type 4 tumor where evaluation of constipation, urine retention, and lower-limb paresis incidentally picked up an abdominopelvic tumor. Here, the sacrococcygeal mass was revealed by per-rectal examination and its extent was defined using image investigations. Tumor markers (AFP and beta-HCG) should be done to diagnose the malignant nature of the tumor. The AFP levels should be interpreted carefully considering different levels of AFP at infancy. Imaging investigations (chest, abdomen, and pelvis) should be done to define the extent of the tumor including intraspinal extension and to see for any distant metastases.

Garg et al. reported five cases of malignant SCT that presented with intraspinal extension. All children were older than 12 months, and 80% of them had neurologic symptoms including lower-limb paresis and bladder or bowel incontinence at the time of presentation. These cases were managed by giving neoadjuvant chemotherapy to shrink the lesion followed by complete surgical resection. This option has excellent results in decreasing the tumor size and diminishing the need for spinal exploration. Garg et al. series had all cases beyond infancy, thereby alluding to the natural course of the sacrococcygeal teratomas wherein the tumor grows into the intraspinal area.

Very similar to the aforementioned series of Garg et al., in our children also, neoadjuvant chemotherapy achieved a good response resulting in shrinkage of the tumor, disappearance of lung metastases, and disappearance of intraspinal component making surgical excision easier without surgery-related morbidity. We concur with Garg et al. that the resolution of neurological symptoms post treatment suggests that the neurological symptoms are due to compression of the nerves by the intraspinal component rather than infiltration and destruction of nerves. Our study also supports neoadjuvant chemotherapy as the first-line modality for all children of SC-MGCT with intraspinal extension. This makes the tumor easily resectable without the need vertebral resection or intraspinal exploration.

**Conclusions**

Malignant sacrococcygeal GCTs with intraspinal extension are rare and can be managed with neoadjuvant chemotherapy which obviates the need for intraspinal exploration and has got good neurological and oncological prognosis.
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Conflicts of interest
There are no conflicts of interest.

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