Sex cord-stromal tumors (SCST) of ovary comprise around 5% of ovarian neoplasms. Ovarian fibromas are one of the most common benign SCST of the ovary usually seen most commonly in the 5th decade of life and rarely in the pediatric age group. We operated on an 18-month-old toddler; she presented with abdominal distension and on evaluation was diagnosed as having ovarian mass with ascites. Intraoperatively right ovarian mass with ascites noted seen and laparoscopy-assisted right salpingo-oophorectomy done. Final histopathology was reported as an ovarian fibroma.

Keywords
Sex cord stromal tumors; Ovary; Fibroma

1. Introduction
Sex cord-stromal tumors of ovary comprise around 5% of ovarian neoplasms. They arise from the sex cords or the ovarian stromal cells. SCST are primarily classified into pure stromal tumors, pure sex-cord tumors and mixed sex-cord stromal tumors of which fibroma is the most common benign SCST [1]. Ovarian fibromas are one of the most common benign SCST of the ovary usually seen in the 5th decade of life and rarely in the pediatric age group [2]. To the best of our knowledge, a total of 13 ovarian fibroma cases have been reported in the pediatric and adolescent age groups in world literature till date (Table 1) [2]. We report a case of unilateral ovarian fibroma of the right ovary in an 18-month-old toddler along with a brief review of the literature.

2. Case report
An 18-month-old toddler was investigated for gradually progressing abdominal distension of 10 days duration. Contrast-enhanced computed tomography abdomen showed a well-defined, dumbbell-shaped lobulated soft-tissue attenuation lesion. The lesion measured 80 × 44 mm, was located in the supravesical region extending to right iliac fossa. It showed few cystic areas with faint enhancement on contrast study, located in the supravesical region extending to right iliac fossa. The lesion was drained by the right ovarian vein (likely right ovarian neoplasm). The child was born through a non-consanguineous marriage and delivered by cesarean section. The baby was breastfed and had no congenital defects. There was no family history of breast or ovarian malignancy. Tumor markers, HCG: < 0.100 mIU/mL; LDH: 207 U/L; Serum AFP: 3.12 ng/mL; CA-125: 3 U/mL all were within the normal range. Her parents were counselled, and she was planned for surgery.

Intraoperatively, 750 mL free fluid in the peritoneal cavity was drained and a 7 × 4 cms dumbbell-shaped mass seen arising from the right ovary (Fig. 1). Ascitic fluid was collected for cytology. Laparoscopy-assisted right salpingo-oophorectomy was done, and specimen sent for frozen section. Frozen section was suggestive of a sex-cord stromal tumor with differentials of juvenile granulosa cell tumor or ovarian fibroma. Postoperatively she recovered well and was discharged on the 4th post-operative day. During the minimal access procedure, we ligated the right infundibulopelvic ligament and right tubo-ovarian ligaments with a vessel sealer. The specimen was held by the cut edge of the infundibulopelvic ligament and placed in an endobag. These maneuvers avoid tumor handling.

Histopathological examination of the right salpingo-oophorectomy specimen showed a 5.9 × 3.6 × 2.7 cm solid homogenous tumor weighing 65 g and replacing the entire right ovary. The cut section of the specimen shows ovarian parenchyma harboring a lesion arranged in diffuse sheets, trabeculae, and microcystic architecture. Lesional cells showed indistinct cell borders with scant eosinophilic cytoplasm and elongated to oval mildly pleomorphic vesicular nuclei. Occasional mitoses were noted, and scattered thin-walled blood vessels were seen. Immunohistochemistry was positive for Vimentin and negative for Inhibin, Calretinin, Glypican 3, CD10, WT1, Beta-catenin, Cyclin D1, and TLI1, suggestive of ovarian fibroma with microcystic change (Fig. 2).

3. Discussion
Ovarian fibroma is the most common benign ovarian sex cord-stromal tumor [1]. It comprises 5% of all SCSTs. Fibromas, though seen in all age groups, are most common in postmenopausal women and are hormonally inactive tumors. Our patient was 18-months-old. Review of the medi-
Fig. 1. Laparoscopic view of the ovarian fibroma. (a) Ascites. (b) Image showing right ovarian mass and normal left ovary. (c) Normal uterus and both fallopian tubes. (d) Picture showing resection of the right ovarian mass.

<table>
<thead>
<tr>
<th>S. No</th>
<th>Author</th>
<th>Age</th>
<th>Unilateral or bilateral</th>
<th>Size of the tumor (greatest diameter)</th>
<th>Other associated findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Charache et al.</td>
<td>3 years</td>
<td>Unilateral</td>
<td>NA</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>Bower et al.</td>
<td>5 years</td>
<td>Bilateral</td>
<td>NA</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>Rater et al.</td>
<td>5 years</td>
<td>Unilateral</td>
<td>NA</td>
<td>Gorlin’s syndrome</td>
</tr>
<tr>
<td>4</td>
<td>Junaid et al.</td>
<td>3 years</td>
<td>Unilateral</td>
<td>NA</td>
<td>Multiple cutaneous meningiomas</td>
</tr>
<tr>
<td>5</td>
<td>Dumont-Herskowitz et al.</td>
<td>3 years</td>
<td>Unilateral</td>
<td>5 cm</td>
<td>Familial</td>
</tr>
<tr>
<td>6</td>
<td>Dumont-Herskowitz et al.</td>
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<td>Unilateral</td>
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</tr>
<tr>
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<td>NA</td>
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<tr>
<td>9</td>
<td>Raggio et al.</td>
<td>7 years</td>
<td>Bilateral</td>
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<td>Gorlin’s syndrome</td>
</tr>
<tr>
<td>10</td>
<td>Johnson et al.</td>
<td>3.5 years</td>
<td>Bilateral</td>
<td>8 cm in right ovary; 4 cm in left ovary</td>
<td>Gorlin’s syndrome</td>
</tr>
<tr>
<td>11</td>
<td>Howell et al.</td>
<td>8 years</td>
<td>Bilateral</td>
<td>9 cm in right ovary; 6 cm in left ovary</td>
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<tr>
<td>12</td>
<td>Laufer et al.</td>
<td>12 years</td>
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<td>None</td>
</tr>
<tr>
<td>13</td>
<td>Chen YJ et al.</td>
<td>7 months</td>
<td>Unilateral</td>
<td>7 cm</td>
<td>None</td>
</tr>
<tr>
<td>14</td>
<td>Present case</td>
<td>18 months</td>
<td>Unilateral</td>
<td>5.9 cm</td>
<td>None</td>
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</table>
Fig. 2. Histomicrograph and IHC pictures of ovarian fibroma. (a) H&E 40× showing circumscribed lesion composed of spindle cells. (b) H&E 400× showing spindle cells in short fascicles with minimal cytological atypia. (c) H&E 400× shows areas with microcystic pattern. (d) IHC 400× shows cells that are positive for vimentin.

Clinical literature reveals a case report of a 7-month-old infant [2]. Ovarian fibromas in the young age group are rarely associated with Gorlin syndrome or Nevoid basal cell carcinoma syndrome [3]. It is an autosomal dominant disorder characterized more commonly by PTCH1 gene mutations, i.e., 9q22.3 microdeletion [4] and less commonly SUFU gene mutations though ovarian fibromas were most common with the later [5]. Gorlin syndrome is also associated with basal cell carcinoma skin, this typically begins to develop during adolescence or early adulthood, keratocystic odontogenic tumors, medulloblastoma, small depressions (pits) in the skin of the palms of the hands and soles of the feet, macrocephaly with a prominent forehead and skeletal abnormalities involving the spine, ribs, or skull. There is no family history suggestive of Gorlin syndrome in our patient, she is too young to develop any features of Gorlin syndrome.

Ovarian fibromas are stromal tumors composed of spindle, oval or round cells that produce variable amounts of collagen. They present as an abdominal lump or with ascites. There are no characteristic signs and symptoms that can preoperatively help in distinguishing ovarian fibromas from other ovarian tumors. Meig’s syndrome characterized by the triad of ovarian fibroma, ascites, and pleural effusion, is seen in around 1% cases only [6]. Serum CA-125 is not a useful biomarker in diagnosing ovarian fibroma as patients may have elevated values which depends on various other factors like tumor diameter, presence of ascites, hydrothorax, or Meigs syndrome [7]. Based on history, physical examination, and imaging, ovarian fibroma cannot be distinguished from other ovarian or solid adnexal tumors, hence it is always diagnosed postoperatively on histopathology [8]. Management options in case of ovarian fibroma include cystectomy, unilateral, or bilateral salpingo-oophorectomy considering the patient’s age, family status, and patient interests. Surgery can be done either by laparotomy or by minimal access based on the expertise [9]. Laparoscopic surgery has several advantages including preservation of ovarian tissue, lower rate of adhesions; a very important factor in young nulliparous patients (for preventing infertility due to tubal factor), shorter hospital stay, faster normalization of bowel activities, faster re-
turn to social life, less morbidity and better cosmetic results in terms of the scar. A conventional staging laparotomy needs a midline vertical incision whereas we finished the procedure with three 5 mm ports and a 10 mm camera port. The specimen was retrieved in an endo-bag through a 6 cms transverse suprapubic incision [10].

Histopathologically fibromas consist of spindle cells in a whorled arrangement with eosinophilic cytoplasm, fusiform nuclei, and a variable amount of extracellular collagen. Cytologic atypia is rare. These fibromas must be distinguished from other similar rare entities like cellular fibromas 1-3 mitoses per 10 HPF, mitotically active cellular fibromas ≥ 4 mitoses per 10 HPF or fibrosarcomas with moderate to severe atypia and elevated mitotic rates [11, 12].

4. Conclusions

Ovarian fibromas in the first decade of life are extremely uncommon. As discussed, an extensive literature search has revealed only one patient younger than this toddler. We successfully completed the procedure through laparoscopy, and the specimen was bagged and retrieved through a small suprapubic incision. The toddler had an uneventful postoperative recovery and was discharged on the 4th postoperative day. The advantage of a minimal access procedure cannot be overemphasized especially in toddlers and infants. It reduces the abdominal incision and retraction, thus reducing postoperative pain and facilitates an early discharge.

Author contributions

All authors contributed equally in the development of the manuscript.

Ethics approval and consent to participate

Written informed consent and ethics committee approval was obtained from the patient’s family for publication of this case report and accompanying images.

Acknowledgment

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Conflict of interest

The authors declare no conflict of interests.

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

References