CASE 2

DIAGNOSTIC ENIGMA:MULLERIAN TISSUE IN SPINAL CORD

Dr. Saloni Bharadwaj
Resident, Department of Pathology

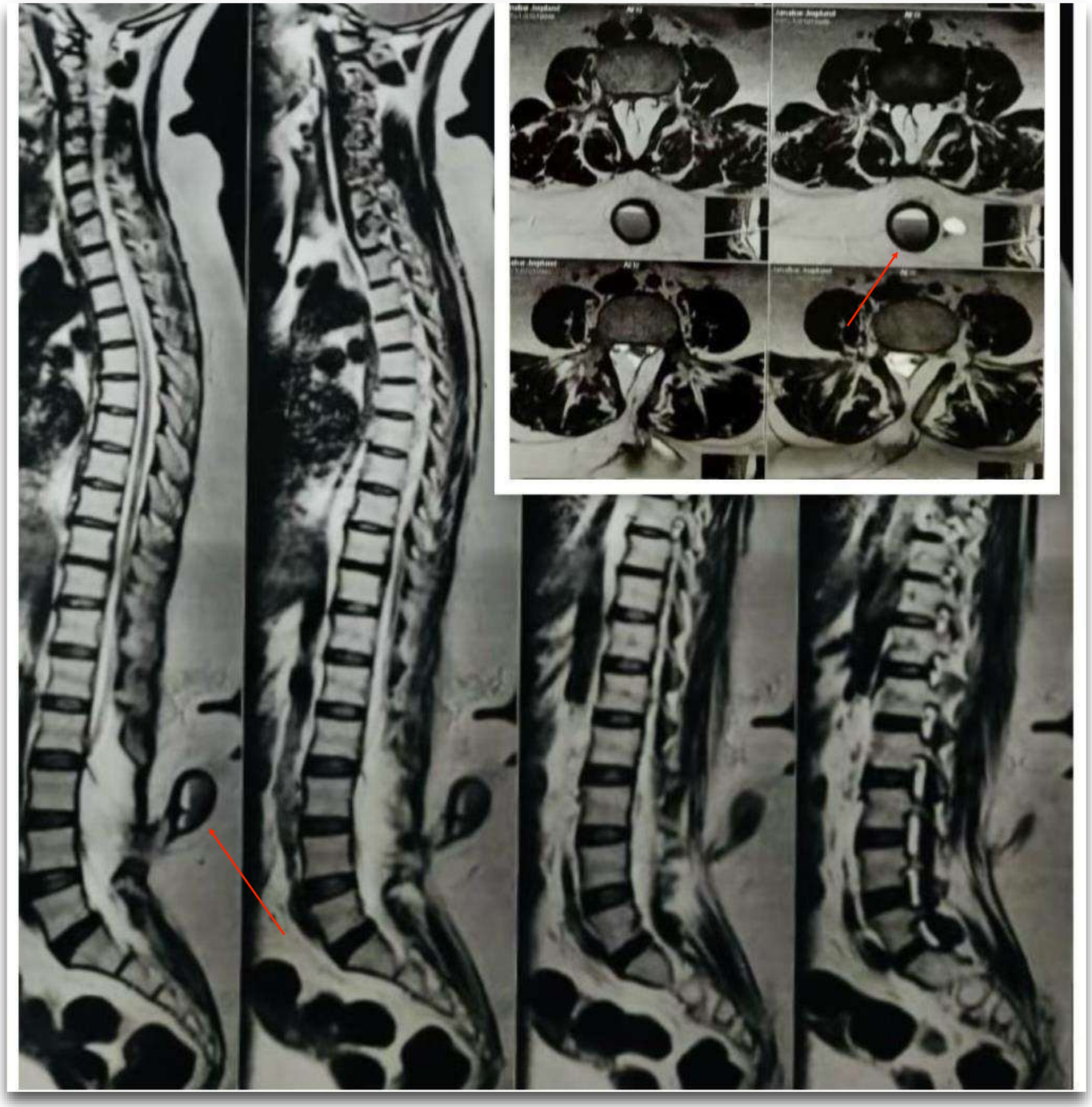
Clinical details

- 39 yrs/ Female presented to Neurosurgery OPD
- Lower back pain since >10 yrs, radiating to bilateral lower limb.
- Tingling and numbness in bilateral lower limb since 6 months.
- Difficulty in walking since 6 months.
- Increased frequency of urine.
- No prior history of trauma/ swelling.
- All the routine haematological and biochemical parameters- Within normal limits.
- No h/o weight loss/ loss of appetite/ bowel disturbances.
- No h/o DM/ HTN/ TB/ Asthma/ relevant surgeries/ addictions.

MRI Lumbosacral spine:

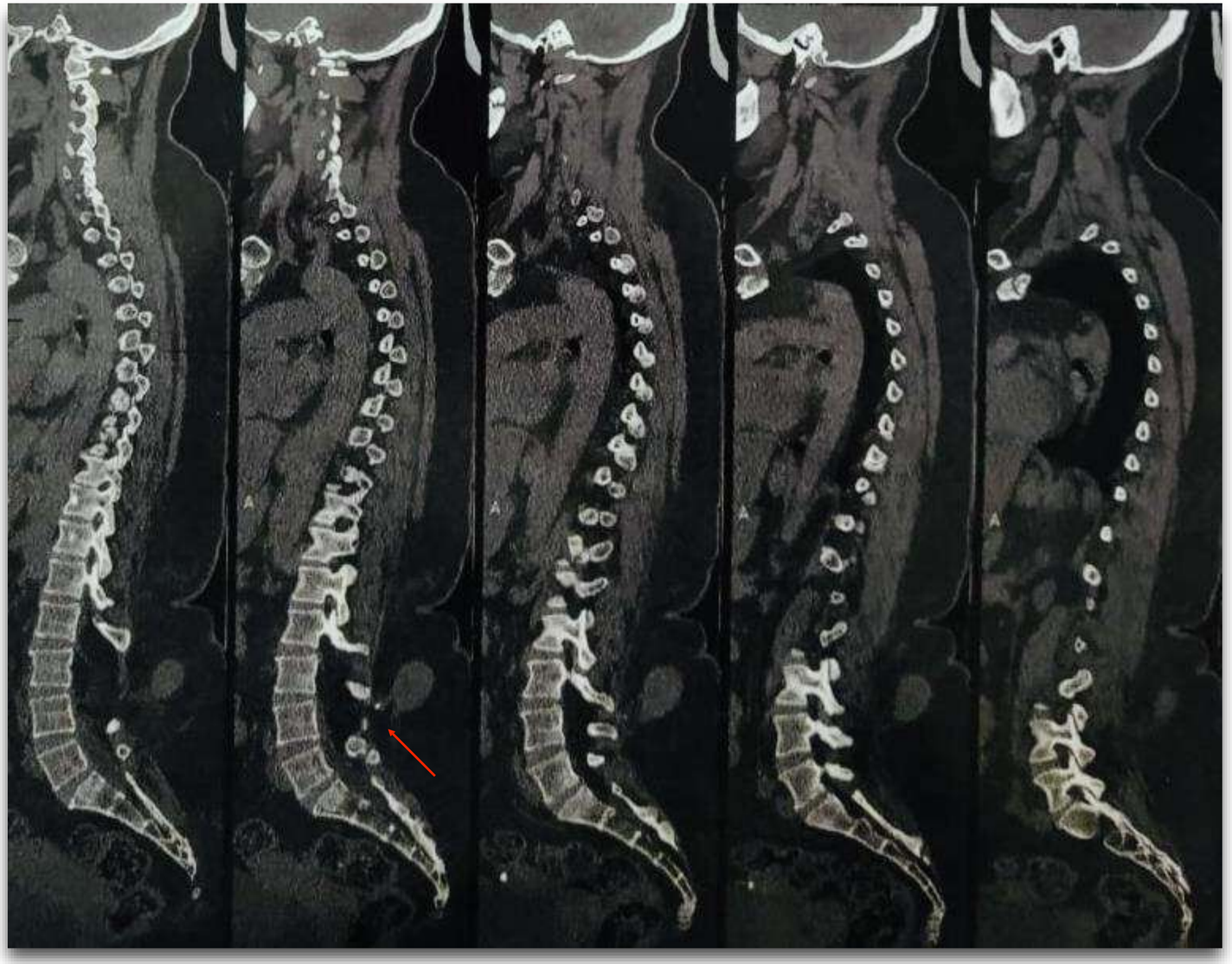
- Suggestive of spinal dysraphism- **spina bifida at L3-L4 level and lipomyelocele**.

- A well-defined thick walled lesion within subcutaneous fat (3.1 x 1.9 x 2.3 cm) showing predominant fat component with fat fluid level along the anterior aspect - likely suggestive of **dermoid cyst**.



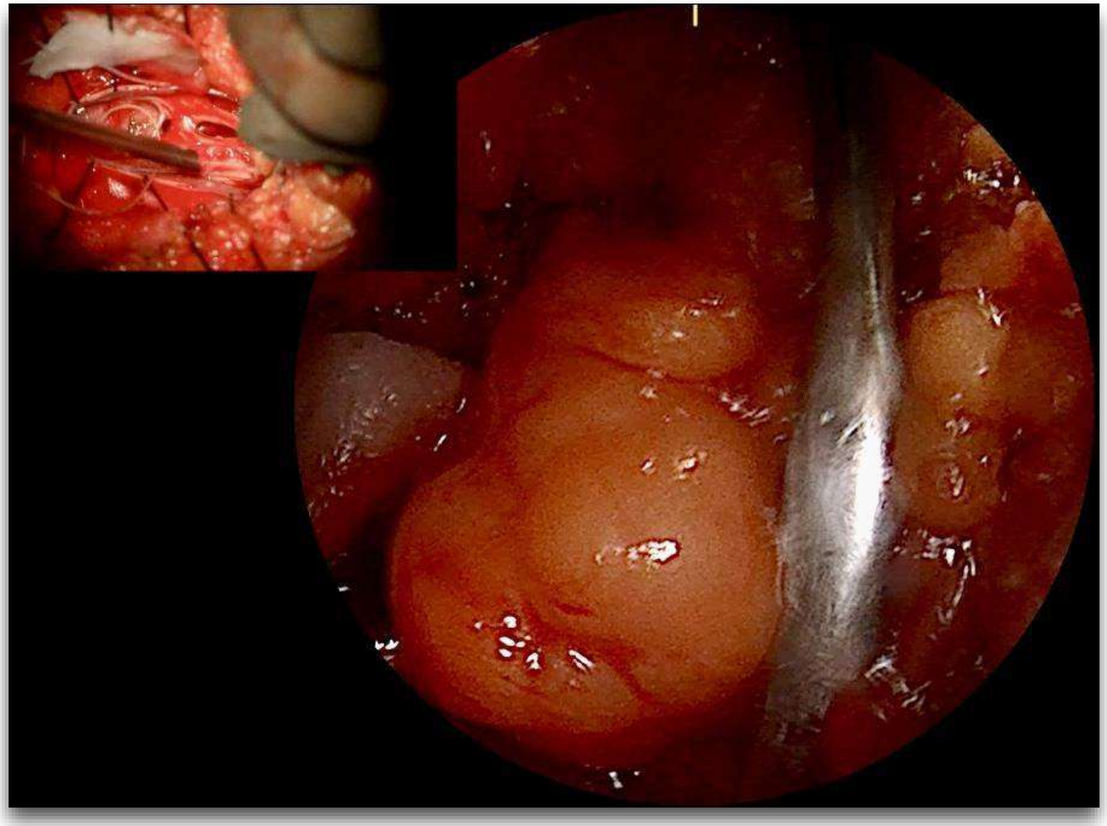
CT WHOLE SPINE (PLAIN):

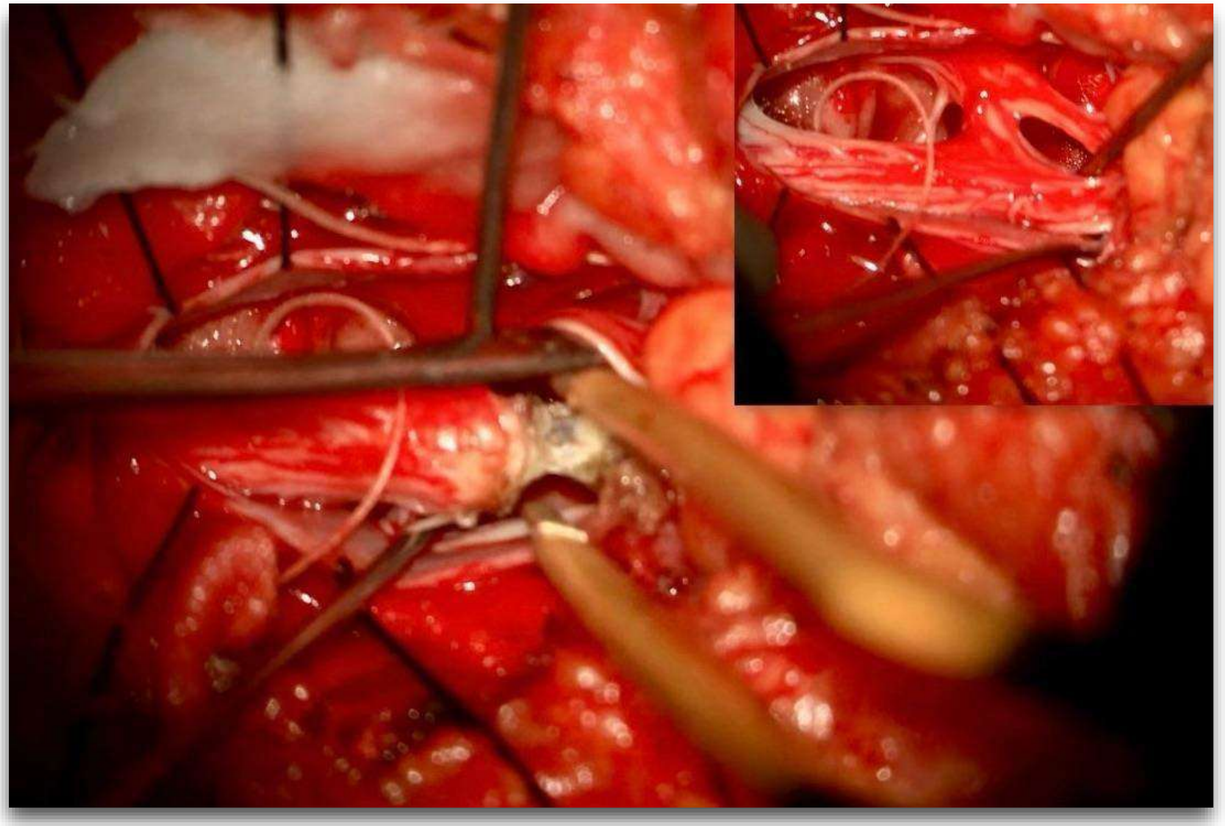
- Suggestive of **Spinal dysraphism-spina bifida at L3-L4 level with lipomyelocele.**



Surgical intervention

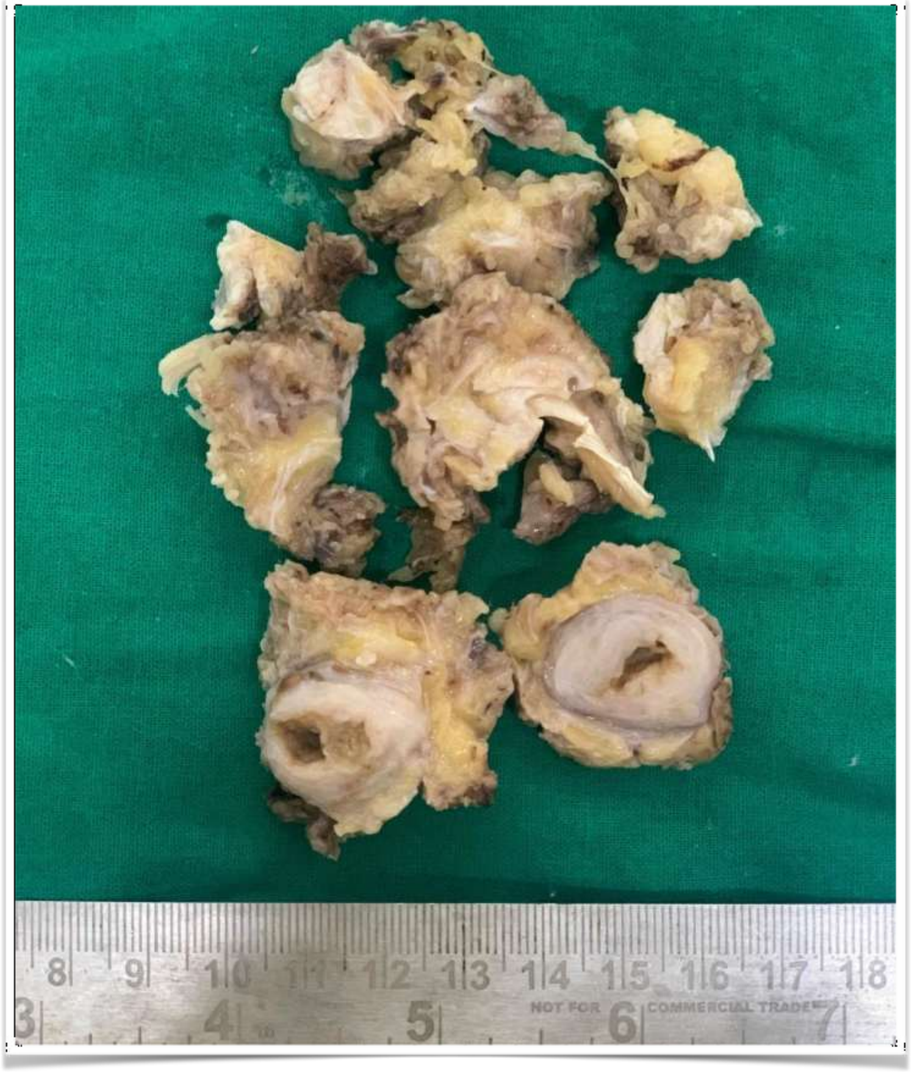
- Laminectomy from level L2-L5 with repair of lipomyelocele and excision of the dermoid cyst with detethering of cord.



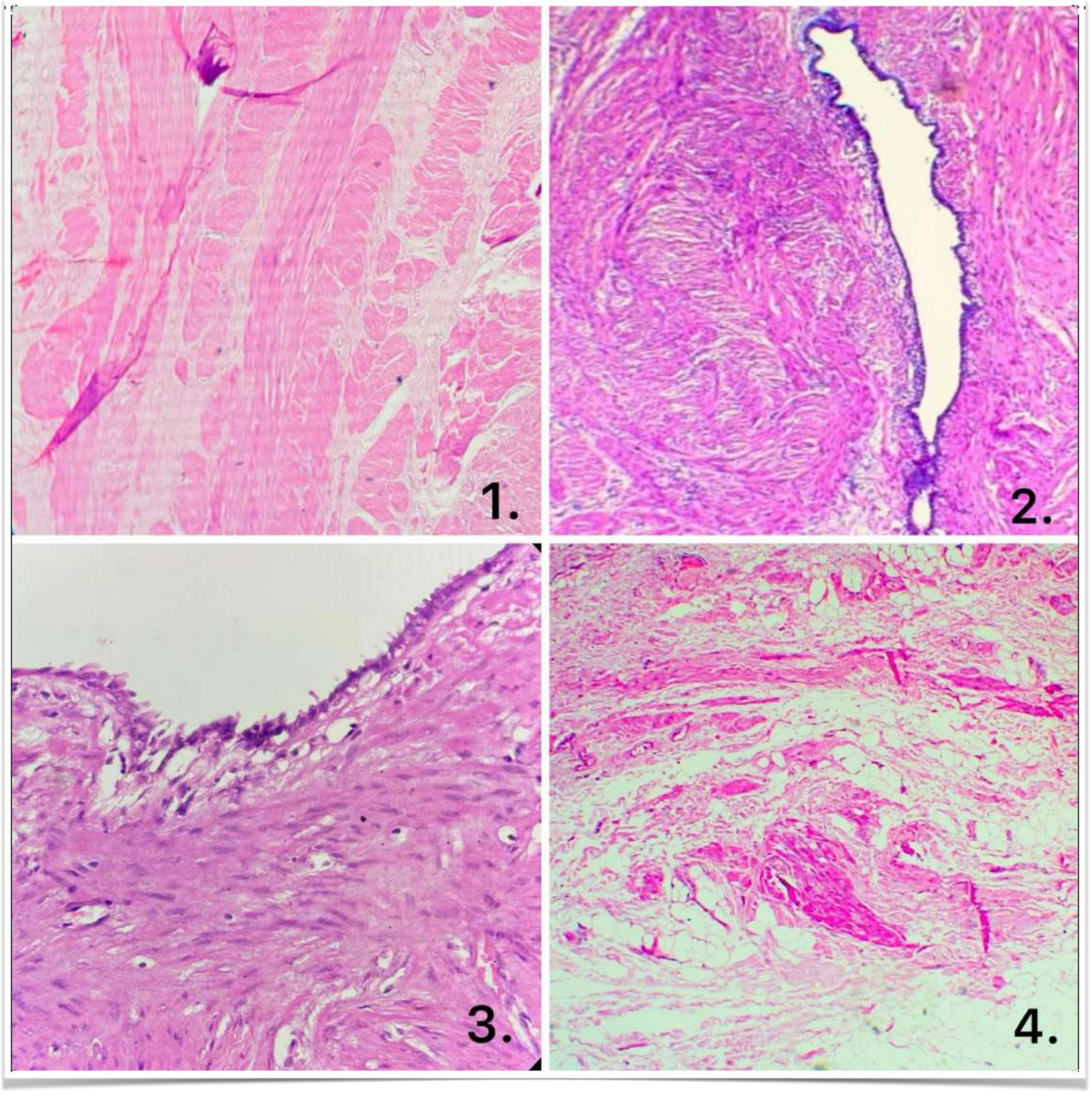


Gross description

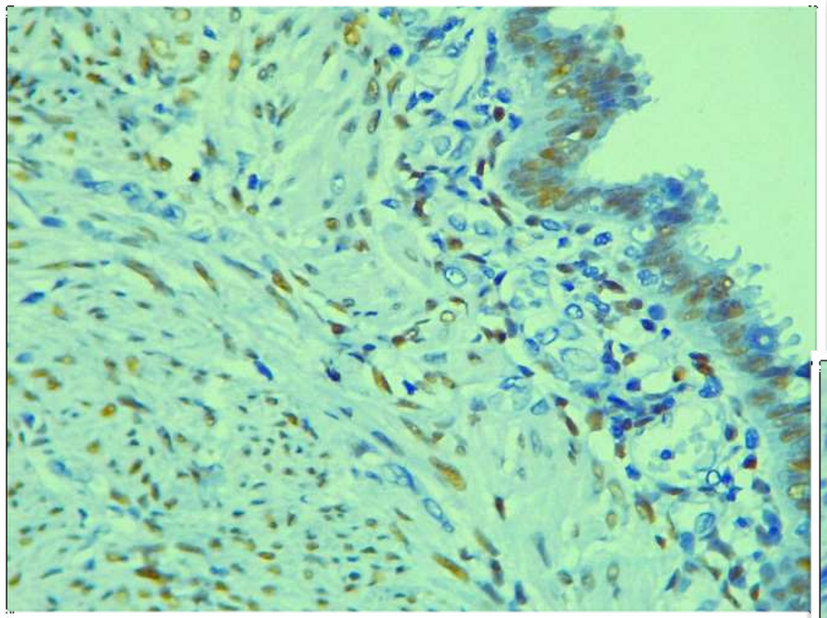
- Received multiple fibro-fatty, soft to firm tissue pieces, aggregating to 6.5 x 4.5 x 3cm.
- On cutting, one of the tissue pieces showed thickened wall with an opening measuring 2 x 1.8cm, resembling uterus.



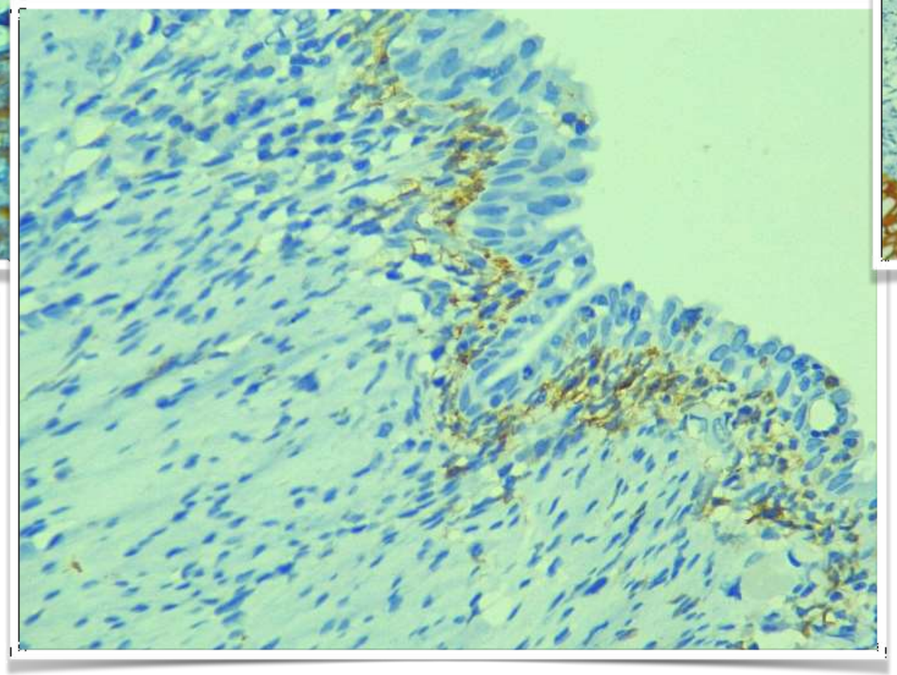
1. Thick smooth muscle layer resembling myometrium.
2. Lining of endometrium
3. Ciliated cell metaplasia
4. Mature adipocytes with nerve bundles



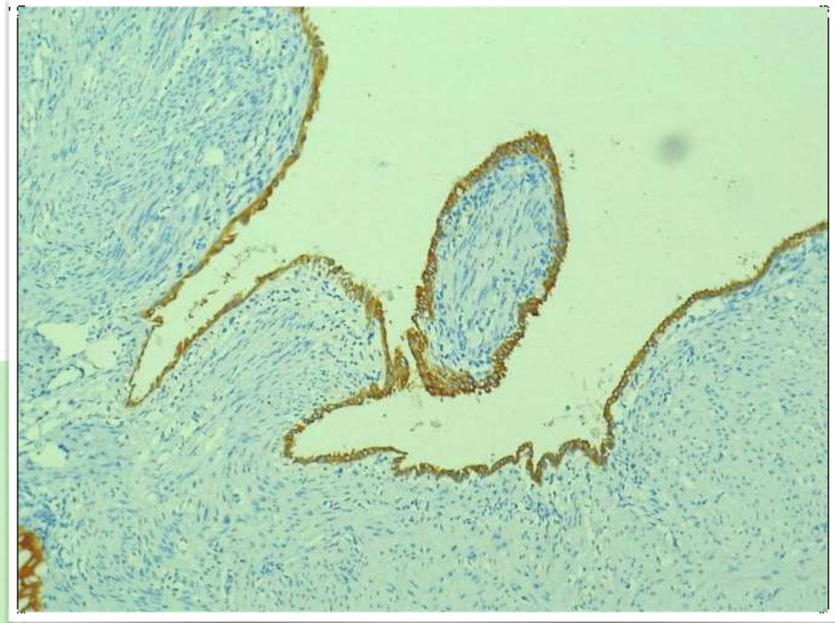
IMMUNOHISTOCHEMISTRY



ER: Positivity in epithelial, stromal and muscle cells.



CD-10: Positive in sub-epithelial stromal cells.



CK-7: Positive in epithelium

Histopathology Diagnosis - Mullerian Choriostoma with lipomyelocele.

Dermoid cyst ruled out - No evidence of ectodermal elements.

The patient recovered well after the surgery. The symptoms showed significant improvement on **follow-up visits**.

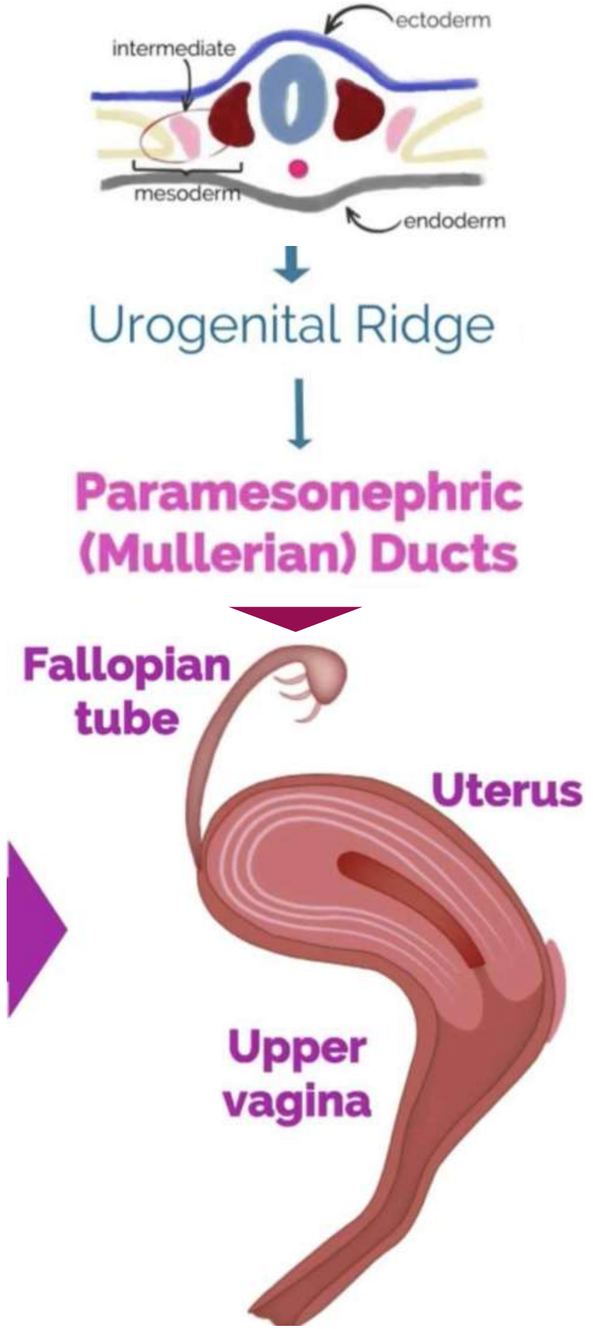
DISCUSSION

- **Choriostomas-** cohesive tumor like mass consisting of normal cells in an abnormal location.
- Rare to find mullerian choriostomas with spinal dysraphism, <10 cases published in literature, age ranged from 4 days to 61 yrs old females.
- Described in diverse locations- the spinal cord, ureter, urinary bladder, sciatic nerve etc.
- Usually found in the lumbo-sacral region- composed of endometrial, endosalpingeal and endocervical tissues.
- IHC shows positivity for ER, CD10 and CK7.

- **Etiology not fully understood**- one hypothesis is that they result from misplacement of Mullerian duct tissue during embryonic development.

- Development of the female reproductive system is regulated by various homeobox genes (Hox, Nkx, Phox).

- Also responsible for the development of normal mullerian system at heterotopic sites leading to the formation of mullerian choriostomas.



- **Differential diagnosis** includes endometriosis, hamartoma, and teratoma.
  - Endometriosis - does not show organoid formations with smooth muscle.
  - Hamartoma- shows disorganised growth of mature tissues normally present in the affected part.
  - Teratoma- composed of a heterogenous mixture of mature or immature tissues, derived from >1 germ cell layer.

Since our case displayed an organised, well-formed uterine tissue and did not contain any e

References

1. Kakkar A, Sharma MC, Garg A, Goyal N, Suri V, Sarkar C, et al Uterus-like mass in association with neural tube defect: A case report and review of the literature Pediatr Neurosurg. 2012;48:240–4
2. Batt RE, Yeh J. Müllerianosis: Four developmental (embryonic) mullerian diseases Reprod Sci. 2013;20:1030–7
3. Christodoulidis G, Zacharoulis D, Barbanis S, Katsogridakis E, Hatzitheofilou K. Heterotopic pancreas in the stomach: A case report and literature review World J Gastroenterol. 2007;13:6098–100

Thank you!