The chest X ray PA views was normal. The serology was non-reactive. The CT-Abdomen was normal. The MRI-Abdomen showed a 1 cm Urachal Cyst.

The final pre-op diagnosis was: Acute Appendicitis and Remnant Urachal Cyst Laparoscopic Urachal cyst excision and Appendectomy was done and sent for Histopathological examination. Preoperatively, the appendix appeared grossly unremarkable.

Histopathology Study showed: Urachal cyst excised specimen showed a cyst lined by transitional epithelium (Figure1).

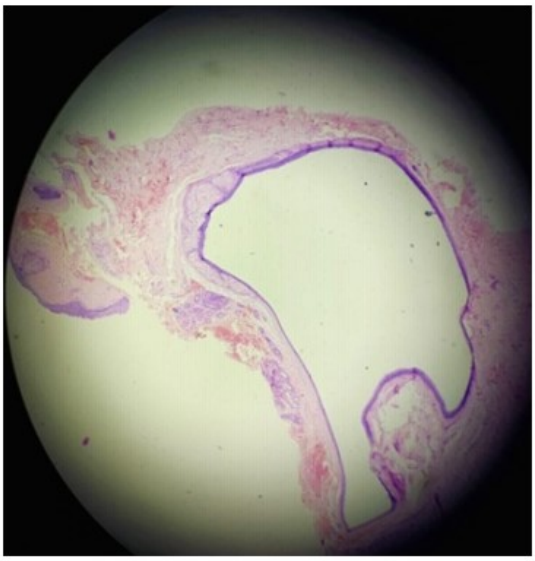

Figure 1: Haematoxylin and eosin stained section (10Xx4X) showing urachal cyst lined by transitional epithelium.

Appendicectomy specimen showed a Goblet Cell Carcinoid. Grossly, the appendix appeared shrivelled up with a yellowish tip measuring 1X1 cm (Figure 2).

Figure 2: Gross appearance of the appendix with a yellow tip.

Three sections were taken one each from the tip; mid and base of the appendix. Tumour was seen at the tip with circumferential involvement. The tumour was composed of nests, trabaculae and cords of atypical cells separated by extra-cellular pools of mucin. The atypical cells showed mild atypia, a vesicular nucleus and "salt and pepper" chromatin and eosinophilic vacuolar cytoplasm. The tumour was seen invading the muscularis layer and extending up to the serosa and was not seen infiltrating the periappendiceal fat. The tumour showed characteristic nesting pattern at the periphery of the tumour. The tumour showed both intra and extra cellular pools of mucin (Figure 3).

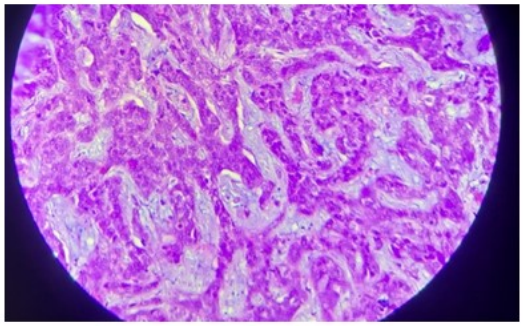

Figure 3: Haemaoxylin and eosin stained section (10XX40X) showing intra and extracellular pools of mucin.

At the periphery of the lesion some desmoplastic was seen. Mitotic Figures 3-4/10 hpf was seen. Paneth cell metaplasia was seen. The immunohistochemistry showed cells to be Chromogranin diffusely positive (Figure 4) and Ki 67 index was 3-4%.

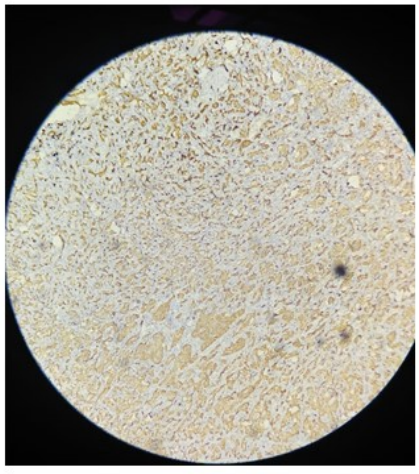

Figure 4: Immunohistochemistry with Chromogranin (10XX10X) showing diffuse Chromogranin positivity.

Thus, the tumour was classified as "Goblet cell carcinoid" Grade II pT_3 since there was no breach of the serosa. Any perineural or vascular invasion or periappendiceal involvement of the appendix by the tumour was not seen as is commonly seen in these tumours [2].

DISCUSSION

The age at presentation of Goblet cell carcinoid is unusual in our case (18 years) [3,4]. It has been reported to occur in an older age group with equal predilection of the sexes. Half the female patients presented with metastasis to the ovary [5]. Our patient's ovary was free of disease.

Goblet cell carcinoid can present with pain in right iliac fossa as in our case [6-8]. Incidental finding of goblet cell carcinoid is seen in 3% cases [5]. Presentation with stage III/IV disease can be seen in 51% to 97% [8,9], cases as was seen in our patient. Lymph node metastasis was seen in 17-38% cases with involvement of the mesenteric node [5,8]. The patient did not have carcinoid syndrome [9,10].

These tumour is generally seen at the tip of the appendix as an ill-defined mass with circumferential and longitudinal extension as was seen in our case [5]. It may involve the whole of the appendix which was not seen in our case where the tumour was confined to the tip. The mucosa was spared as was seen in our case.

Microscopically, goblet cell carcinoid has been divided into Grade I, II and III. Grade I being cells with minimal atypia, mitotic figures <2/hpf and Ki 67 index<2%. Grade II having cells with moderate atypia, mitotic figures between 2-20/hpf, Ki 67 index 3-20% and Grade III being cells showing marked atypia with mitotic figures >20/hpf and Ki67 index>20% [10]. Our patient had Grade II tumour.

This tumour's generally show strong positivity for CEA, CDX-2 CAM 5.2 and CK. They are inconsistently positive for neuroendocrine markers [6,12], though the tumour in our case showed diffuse Chromogranin positivity [5].

A urachal cyst is a collection of tissue and fluid between the bladder and the umbilicus. The cyst forms in the remnants of the urachus, a structure normally present in a fetus that usually closes before birth. In some infants, the closure doesn't occur.

The urachal cyst remains largely asymptomatic unless infected. Infected urachal cysts present with acute symptoms such as suprapubic pain, dysuria, fever, nausea, vomiting, haematuria, pelvic pain, and purulent umbilical discharge like it did in our patient who had haematuria and repeated urinary tract infection [2].

On post-operative follow-up the patient had gained 3 kg weight and did not have any abdominal mass/pain [12], as the tumour was confined to the appendix.

CONCLUSION

It is important to submit all appendixes for histopathological examination where especially the tip of the appendix should be examined.

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