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Case Report

Primary carcinoid tumour of the testis

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INTRODUCTION

Testicular carcinoid tumours are very rare and account for less than 1% of all testicular neoplasms.[1] These tumours may be classified into three distinct groups, most commonly (1) primary testicular carcinoid, (2) carcinoid differentiation within a mature teratoma, and (3) metastases from an extra-testicular source. Testicular carcinoid tumours do not follow the age category of men affected most commonly by germ cell tumours (20-40 years), cases have been reported ranging in age from ten to eighty-three years.[2] Presentation of carcinoid tumours may be with self-detected testicular mass or testicular ache as with common testicular tumours, or uncommonly with carcinoid syndrome.

We report a case of primary carcinoid tumour of the testis without features of carcinoid syndrome.

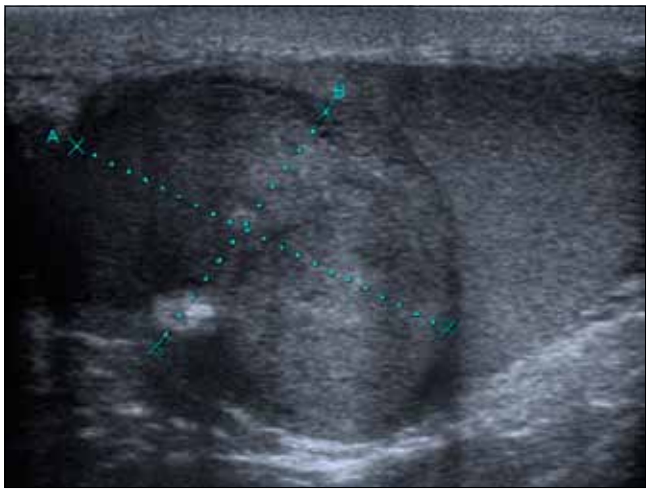

Fig 1. Ultrasound image of the right testis showing mixed echogenic mass in upper pole in keeping with testicular tumour

CASE REPORT

A 29-year-old male presented to the outpatient clinic with a several week history of a painless right sided testicular swelling found on self-examination. He had no recent history of trauma, urinary tract or sexually transmitted infections. On examination he had a hard mass in the upper aspect of his right testis which felt strongly suggestive of testicular tumour. An ultrasound scan showed normal appearance of the left testis and a mixed echogenic mass occupying the majority of the superior aspect of the right testis. The mass measured 3.5 x 2.3 cm and had increased vascularity (Figure 1). Tumour markers (beta human chorionic gonadotrophin, alfa-fetoprotein and lactate dehydrogenase) were normal and a pre-operative chest radiograph did not reveal any metastases. The patient underwent radical orchidectomy and insertion of testicular prosthesis soon after diagnosis.

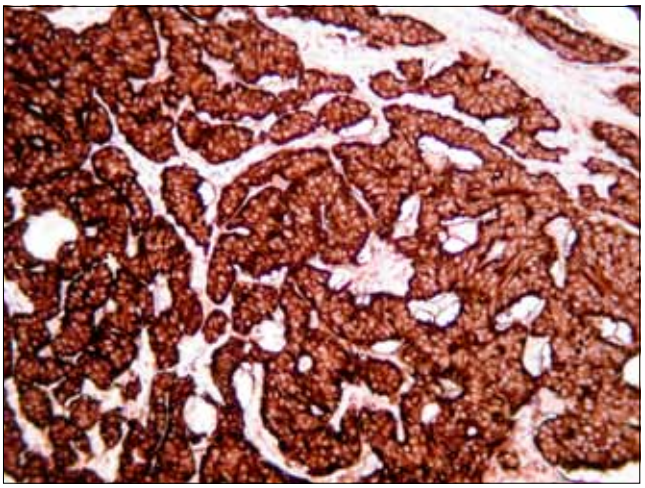

Fig 2. Monomorphic cells in nested trabecular pattern. Staining strongly positive with chromogranin

Histological analysis showed a well circumscribed tumour composed of monomorphic cells arranged in a nested trabecular pattern. The tumour cells had granular chromatin and scarce mitotic figures. Immunohistochemistry showed strong positive staining with chromogranin (Figure 2), synaptophysin (Figure 3), CD56 and PGP9.5. These features were in keeping with a well differentiated carcinoid tumour with no teratomatous components or other germ cell elements. There was no lymphovascular invasion, the tumour was confined to the testis and had a low proliferation index.

The patient subsequently had a staging computed tomogram (CT) of chest, abdomen and pelvis, which showed no significant para-aortic or iliac lymphadenopathy and no pulmonary abnormality. Multiple mesenteric nodes were noted which were not said to be typical of spread of testicular tumour.

The patient was referred to a specialist gastroenterologist for further investigation to rule out the possibility of testicular

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metastasis from an extra-testicular primary carcinoid. NM octreotide scan with SPECT showed two to three subtle foci of activity towards the dome of the right lobe of the liver in segment four and eight. These were not definite for liver metastases, however, these could not be excluded. No lesions were found to be over expressing somatostatin receptors. Further evaluation with MRI confirmed that the areas of abnormality within the liver were consistent with simple cysts.

DISCUSSION

Testicular carcinoid tumours are a rare entity and are almost never suspected preoperatively. The majority are only diagnosed on histopathology as they do not become clinically apparent until there is metastatic spread or the presence of carcinoid syndrome.[2] Carcinoid tumours arise from neuroendocrine cells, however, the presence of neuroendocrine cells has not been described in the testis, leaving the origin of primary testicular carcinoid debatable.[3] Several cellular origins of these tumours have been proposed. Mai et al found that the origin of testicular carcinoid tumours was located in the same progenitor cell from which Leydig cells derive.[4] Merino et al support the possibility of a germ cell origin, finding intra-tubular germ cell neoplasia in the testicular tissue surrounding a pure carcinoid.[3] Thus primary testicular carcinoid may be the remaining component of a burnt out teratoma or due to a one-sided development of teratoma.[3,5]

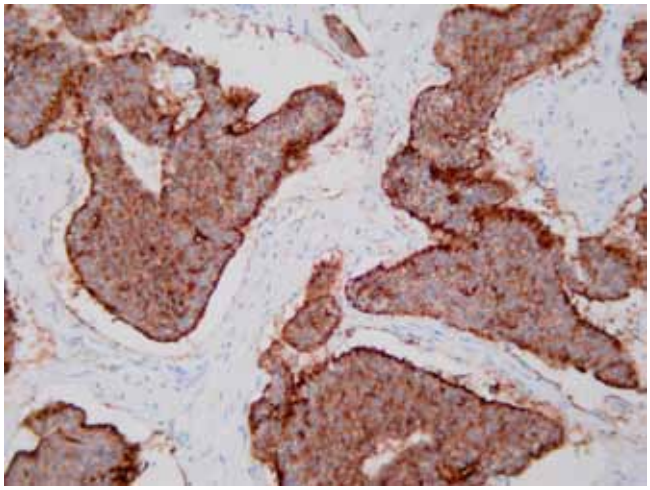

Fig 3. Cells staining strongly positive with synaptophysin

Carcinoid syndrome is manifestation of carcinoid tumours and occurs as a result of the action of vasoactive tumour products. The syndrome is rare, occurring in approximately 10% of patients[5] typically only once the tumour has metastasised to the liver or lungs. Serotonin is the most common tumour product and when released into the systemic circulation it causes the symptoms of carcinoid syndrome.[6] These include increased gastro-intestinal motility, bronchoconstriction, vascular constriction and dilatation.[6] Serotonin is metabolised to 5-hydroxyindoleacetic acid (5-HIAA) which can be measured in the urine. Any patient with vasoactive symptoms and a testicular lump should have 24 hour urinary 5-HIAA performed prior to surgery.

Given that almost 10% of testicular carcinoids are metastases from another location,[2] it is essential to thoroughly investigate these patients to find or exclude an extra-testicular primary source. A multimodal approach has been recommended. Barium contrast studies and CT may detect mucosal thickening or luminal narrowing to suggest bowel involvement. CT is also good for detecting mesenteric extension of the tumour and presence of liver metastases.[7] Somatostatin receptor scintigraphy using indium-111 labelled octreotide is now superior to CT in localisation of primary tumour site and has a sensitivity for detecting metastases of up to 96%.[7] It has superseded meta-iodobenzylguanidine scanning which has a sensitivity of 50% for detecting metastases.[7] These investigations also serve to detect synchronous tumours, as it is known that carcinoids have a high rate of second primary malignancy.[6]

Radical orchidectomy is curative for testicular confined carcinoid.[5] The long-term prognosis of carcinoid tumours is dependent on size, association with teratoma and presence of metastases. Zavala-Pompa et al showed that larger tumours (7.3 vs 2.9cm) and the presence of carcinoid syndrome predicted increased metastatic potential and hence poorer prognosis.[8] The prognosis of carcinoid tumours arising within teratoma is better than pure testicular carcinoid.[8] There have been several reports of carcinoid tumours causing delayed metastases, in one case 17 years after initial diagnosis, highlighting the need for long-term follow-up.[9] Patients should undergo biochemical and radiological follow-up, however, the frequency and duration of follow-up remains open to debate. Sutherland et al suggest that patients should undergo three monthly 5-HIAA measurements for the first year after diagnosis and annually thereafter.[10] More recent guidelines suggest that urinary 5-HIAA levels do not accurately correlate with disease progression and metastases may occur in the absence of an elevated urinary 5-HIAA. Serum chromogranin A (a glycoprotein secreted by carcinoid tumours) has been reported to be a sensitive and specific marker which may correlate with relapse in gastrointestinal carcinoids.[7] It may also be of use in the follow-up of testicular carcinoids, a fast rising level being associated with a poor prognosis.

In conclusion, testicular carcinoid tumours are very rare. It is imperative that once a testicular carcinoid tumour has been diagnosed; the patient undergoes thorough investigation to rule out an extra-testicular primary and metastases. Long-term biochemical and radiological follow-up is essential given potential for delayed metastases.

The authors have no conflict of interests

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