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A case of carcinoid tumour of testis

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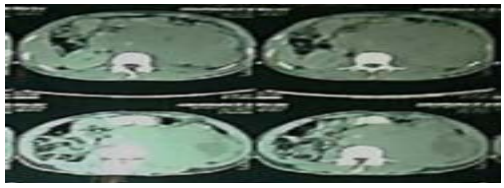
Abstract : Testicular cancers account for around 1-1.5 percent of male neoplasms. A vast majority are germ cell tumours (more than 90 percent) followed by lymphomas (around 5 percent) and interstitial tumours (around 2 percent). Carcinoid tumour of the testis is a rare tumour accounting for less than 1 percent of all testicular tumours. It is usually confined to the testis. Fewer than 100 cases have been reported in the literature.

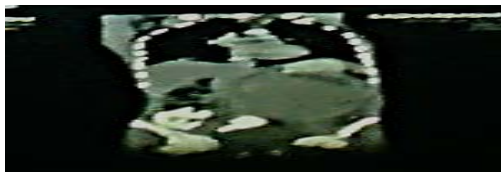
Keyword :carcinoid tumour, testis

CASE REPORT

27 year old male was admitted with complaints of Pain abdomen for 8 months, Mass abdomen for 6 months and Scrotal swelling for 4 months. The abdominal pain was intermittent, dull aching in nature and progressively increasing in severity. The abdominal mass was located on the left side which was initially small and gradually increased in size. He noticed left sided scrotal swelling for 4 months which gradually increased in size. It was not associated with pain or trauma. He had history of loss of appetite and loss of weight. There was no history of fever / chest pain / breathlessness / cough / vomiting / hematemesis / malena / altered bowel habits. On examination, he was thin built and moderately nourished. He was not anemic/ icteric. He had left axillary and bilateral Inguinal lymph nodes which were soft and < 1 cm in size. Abdominal examination revealed a mass of size approximately 15*10 cm palpable involving Left hypochondrium, Left iliac fossa, umbilical and Left lumbar regions which was hard in consistency, had nodular surface, was immobile and not bimanually palpable or ballotable. All borders were well made out. Upper border was 2 cm below costal margin at Left mid-clavicular line, lower border at the level of anterior superior iliac spine, medially extended about 2 cm to the right of midline and laterally 3 cm to the left of Left mid-clavicular line. There was no hepatomegaly or free fluid. Supraclavicular fossa was free. Examination of the scrotum showed 6*4 cm enlarged left testis, which had a smooth surface and was hard in consistency. There was loss of testicular pain sensation on the left side. The provisional diagnosis was left testicular tumour with retroperitoneal lymphadenopathy probably due to seminoma.



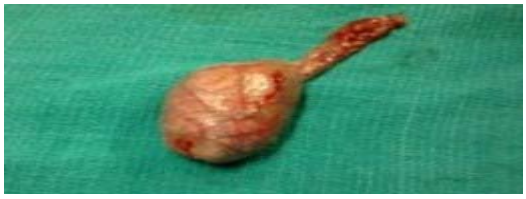
CT abdomen - axial view



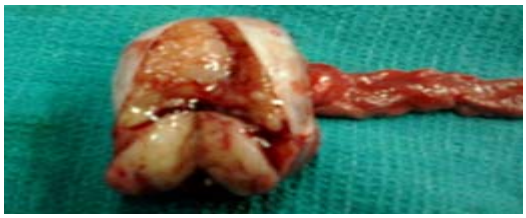
CT abdomen - coronal view

Routine blood investigations namely CBC, RFT and LFT were normal. Tumour markers were done and LDH and HCG levels were elevated [LDH – 1454 U/L; AFP – 1.14 IU/ml; HCG – 71.6 mIU/ml]. Ultrasound scrotum showed an enlarged left testis with multiple loculations with altered echogenicity with mass lesion near lower pole. Ultrasound abdomen showed conglomerate retroperitoneal paraaortic nodal mass 14*7 cm in size. CT abdomen showed a 15*20 cm well defined heterodense lobulated lesion with specks of calcification involving left para-aortic region from L2 to L4 encasing Aorta , IVC , Left Renal vein, coeliac plexus, Superior mesenteric Vein ; compressing upper pole of Left kidney & Pelvi-ureteric junction causing Lt hydronephrosis; infiltrating Lt psoas muscle; small intestine was lifted anteriorly. There was no hepatomegaly and no intestinal abnormalities were noted. The final impression was retroperitoneal necrotic lymph node. CT chest was found to be normal. FNAC of axillary and inguinal lymph nodes showed only inflammatory changes. The patient was planned for high orchidectomy followed by chemotherapy. Left high inguinal orchidectomy was done and a 2 * 2 cm tumour occupying lower

pole of the testis was found. Cut surface had a Cut potato like appearance and was sent for histopathological analysis.



Gross Orchidectomy specimen



Cut section of left testis

HPE report was given as external surface – smooth and cut surface showing multiple grey white nodules largest 2 cm interspersed amidst normal parenchyma. Nodule is grey white with focal yellowish areas showing nests of round to oval cells separated by vascular fibrous stroma; tumour cells are uniform in appearance with coarse chromatin pattern; adjacent Blood vessels show tumour emboli; tubule shows atrophy; no infiltration into epididymis/sac wall; resected end of the cord – normal. The final impression was neuroendocrine well differentiated (carcinoid) tumour of the testis. Immunohistochemistry analysis showed weak positivity for Synaptophysin & Chromogranin and diffuse strong positivity for Neuron specific enolase. The case was discussed in tumour board and was categorised as stage IIIA (T1N3M0S2). He was put on BEP (Bleomycin, Etoposide, Cisplatin) chemotherapy for three cycles and showed more than 50% reduction in size of the nodal mass.

DISCUSSION

Carcinoid tumours of the testis constitute less than one percent of testicular tumours.[1] The first reported case of primary testicular carcinoid was described by Simon et al. in 1954.[2] A painless mass and notable testicular enlargement are the two most common presenting features. It usually affects older males (40-60 years). They can be encountered in 3 clinical settings: as a de novo neuroendocrine neoplasm (most common); as a component of a teratomatous tumour; as a metastatic lesion from extra-testicular primary tumour (least common).[2] The cell of origin is debatable and may arise from the same progenitor cells from which Leydig cells arise4 or may have a germ cell origin.[3] 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 of testis rarely metastasize, with the overall incidence estimated at 11%.5 Features of carcinoid syndrome are seen in <10% of cases5 and typically occur 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 increased gastro-intestinal motility, bronchoconstriction, vascular constriction and dilatation.1 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.1 It is necessary to exclude the presence of a primary tumour in another organ before confirming the diagnosis of a primary testicular carcinoid tumour as around 10% of cases are due to metastasis from another location.[2] A multimodal approach has been recommended. Barium contrast studies and CT abdomen 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. These investigations also serve to detect synchronous tumours, as it is known that carcinoids have a high rate of second primary malignancy. Pathologically, the lesions are described as solid yellow-tan in appearance with an exceedingly firm texture due to striking desmoplasia which is characteristically present. Histologically, the neoplastic cells can form discrete islands, trabeculae, strands, glands, or undifferentiated sheets. Immunohistochemical studies show reactivity to antibodies to cytokeratin AE1 and AE3, Chromogranin-A, neuron specific enolase, Synaptophysin, and CD56.8 High orchidectomy remains the main modality of treatment and is curative if tumour is confined to the testis.5 Chemotherapy is not usually indicated unless there is metastasis. The overall prognosis is good. The long-term prognosis of carcinoid tumours is dependent on size, association with teratoma and presence of metastases. Larger tumour and the presence of carcinoid syndrome predicted increased metastatic potential and hence poorer prognosis. The prognosis of carcinoid tumours arising within teratoma is better than pure testicular carcinoid.9 Long term biochemical and radiological follow-up is essential given potential for delayed metastases.10 Urine 5-HIAA (every 3 months for 1 year and then yearly thereafter)[11] and serum Chromogranin-A levels are used for followup.

CONCLUSION

Carcinoid tumour of the testis is a rare tumour accounting for less than 1% of all testicular tumours. It can occur de novo or it can be present as a component of a teratoma. It usually presents as a painless testicular enlargement. Lymph node metastasis is exceedingly rare. Distant metastasis and thus carcinoid syndrome can occur in 10% of cases. Metastasis from carcinoid tumours at other sites has to be excluded by appropriate investigations before a diagnosis of primary testicular carcinoid tumour is made. High orchidectomy is the mainstay of treatment and Chemotherapy is not indicated unless there is metastasis. Prognosis depends on tumour size, association with teratoma and presence or absence of carcinoid syndrome. Long term biochemical and radiological follow-up is essential due to the potential for delayed metastases.

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