CRYPTORCHIDISM WITH BILATERAL SYNCHRONOUS TESTICULAR GERM CELL TUMOUR

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Abstract

Cryptorchidism or undescended testis is a very common anomaly of the male genitourinary system. It is one of the established risk factors for testicular tumour. The commonest malignancy noted in cryptorchidism is seminoma testis. The presence of bilateral synchronous testicular tumour in cryptorchidism is very rare.

CASE REPORT

A 27 year old male patient came with a complaint of pain abdomen. On examination distention of abdomen with tenderness and hypospadias was appreciated. Ultra sonography of inguino-scrotal region revealed enlarged left testis of 70 x 60 x 55 mm having altered echotexture with multiple isoechoic lesions seen in it. Right side testis was not visualized in scrotal sac, it was present in mid inguinal canal, it was bulky measuring 55x55x50mm. Right testis had heterogenous echotexture with raised vascularity on colour flow. Multiple enlarged inguinal lymph nodes were noted. In USG of both Kidneys, ureters & Bladder both kidneys were normal but multiple heterogenous hypoechoic masses were noted in lesser sac, right subdiaphragmatic location and pelvic cavity. Urinary bladder was nearly empty. Blood investigations were within normal limits. Patient was nonreactive to HIV, HCV and HbsAg.

Computed Tomography (CT) scan of the abdomen showed bilateral bulky testis measuring 5.4x4.6 cm (approx.) on left side and 5x4.6 cm (approx) on right side. Multiple metastatic deposits were present in lesser sac, right hypochondrium and right lobe of liver. Serum human chorionic gonadotropin (HCG) (47.48 mIU/ml) and Alfa feto protein (AFP) (>3000 ng/ml) were elevated. SGOT, SGPT and Serum Alkaline Phosphate level were also raised. A provisional diagnosis of testicular tumour with retroperitoneal and left supraclavicular lymphadenopathy with metastasis to liver was considered. The patient was posted for surgery and

bilateral orchidectomy was performed. Post-operative period was uneventful. On Gross examination, one testis measuring 7 x 5 x 5 cm other measuring 5x4x3 cm. Cut surface of one testis is greyish white and cut surface of other is variegated. Histopathological examination of one testis shows tumour cells having moderate eosinophilic cytoplasm, markedly pleomorphic nuclei with prominent nucleoli. There are foci of yolk sac component with reticular pattern of arrangement of tumour cells, hyaline globules and Schiller-Duval bodies.

Other testis also shows foci of yolk sac component with reticular pattern of arrangement of tumour cells, hyaline globules and Schiller-Duval bodies there were also areas of seminomatous component and areas of necrosis. Histological features are consistent with synchronous bilateral germ cell tumour with one testis having pure yolk sac tumour, other testis having mixed germ cell tumor.

Figure1A:

Figure 1B:

Fig.1A shows shiller duval body at 10x, Fig.1B shows Shiller Duval Body at 40x-tubulopapillary sinusoidal structures with central vascular core and cuboidal to columnar epithelial like cell lining (diagnostic of entity when present) Fig, 2B at 40x

Figure 2B:

Fig,2A seminomatous component showing sheets of relatively uniform tumor cells divided into poorly demarcated lobules by delicate septa with lymphocytes and plasma cells at 10x

Discussion

Testicular tumours are more commonly seen in young adults. The most common variants are GCTs, in which seminoma accounts for 52–56%. The incidence of bilateral GCT is approximately 2%. The risk factors for testicular cancer are mainly cryptorchidism, a family history of testicular cancer, a personal history of testicular cancer and intratubular germ cell neoplasia.¹ Around 80–85% of bilateral testicular tumours are metachronous and 15–20% are synchronous. However, the occurrence of synchronous tumour in cryptorchidism is extremely rare. There are only two cases of synchronous abdominal tumour in bilateral cryptorchidism reported (table 1).^{2,3}

Figure 2A:**Table 1: Comparison between the patients with synchronous testicular tumour in undescended testis**

Author	Age (years)	Presentation	Stage	Tumour markers	Surgery	Histology	Follow-up
Kumar <i>et al</i>	30	Flank pain	T ₁ N ₀ M ₀	LDH-1257	Laparotomy and complete excision	Seminoma poorly differentiated	4 cycles BEP
		AFP-2.02		No recurrence in 8 months			
		βHCG-0.38					
Agarwal <i>et al</i>	23	Heaviness in abdomen, low backache, constipation	T ₁ N ₀ M ₀	LDH- N	Laparotomy and complete excision	Pure seminoma	4 cycles BEP
				AFP- N			No recurrence in 60 months
				βHCG- N			
Hameed <i>et al</i>	28	Lower abdominal pain, abdominal distension	T ₁ N ₀ M ₀	LDH-2890	Laparotomy and complete excision	Pure seminoma	4 cycles BEP
				AFP-1.64			No recurrence in 6 months
				βHCG-0.21			

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AFP, α fetoprotein (ng/mL); BEP, bleomycin, etoposide and cisplatin; β HCG, β human chorionic gonadotropin (mIU/mL); LDH, lactate dehydrogenase (U/L); N, normal; T1N0M0, tumour, node, metastasis.

Cryptorchidism or UDT is characterised by the failure of descent of testis in the scrotum. It is more commonly seen in premature male neonates, with the incidence around 1–4% of full term and 1–45% of preterm male neonates.⁴ The pathogenesis of cryptorchidism remains largely unknown, but is most likely multifactorial, involving genetic and environmental risk factors. Spontaneous descent occurs by 6 months of age in 66% of infants. Cryptorchidism is associated with syndromes such as Down, Klinefelter and prune-belly. It is also seen in conditions such as posterior urethral valve, abdominal wall defects—omphalocele and gastroschisis, and neural tube defects—myelomeningocele, spigelian hernia and cerebral palsy.

There is a 4–6 times increased risk of cancer in the UDT. The contralateral descended testis in a cryptorchid male also has a slightly increased risk of developing malignancy. The mechanism behind the increased risk of malignancy in cryptorchidism is due to the abnormal environment with a high temperature in UDT, which prevents the transformation of gonocytes into stem cells and also arrests subsequent apoptosis. These abnormal gonocytes get accumulated within the seminiferous cords, and undergo mutations. Over a period of time they become the source of carcinoma in situ cells and develop into tumours.⁵ The overall risk of GCT in a cryptorchid testis reduces if orchidopexy is carried out before puberty. The risk of malignancy in UDT reduces after orchidopexy, as the testis is relocated into the scrotum which is a low temperature area, hence promoting normal germ cell development and thereby reducing the probability of low sperm count and malignancy.⁶

The diagnosis of malignancy in UDT is made on the basis of clinical suspicion, radiological imaging and histopathological examination. The management is similar to the testicular tumours in the normally descended testis. The serum markers AFP, β HCG and LDH may be elevated.⁷ Imaging with USG, CECT of the abdomen and pelvis may reveal a large intra-abdominal mass lesion with or without necrosis and

calcification. Imaging studies also help in locating the contralateral UDT. Chest X-ray is carried out to look for lung metastasis. In seminoma, the thoracic metastasis in the absence of retroperitoneal disease or elevated serum tumour markers is uncommon. According to Horan *et al*,⁸ the routine chest CT may be associated with a high rate of false-positive findings, which may complicate subsequent therapy. CT of the thorax should be performed in patients with elevated post-orchietomy levels of serum tumour markers, evidence of metastatic disease by physical examination or abdominopelvic CT or abnormal or equivocal findings on chest radiography. The most common histopathological variant of tumour in UDT is seminoma followed by embryonal carcinoma, teratoma and choriocarcinoma.

The treatment of choice for synchronous bilateral testicular tumour is bilateral high inguinal orchietomy. The role of CT-guided/USG-guided biopsy from the testicular tumour is confined to patients with a suspicion of lymphoma or metastasis in the testis.¹ Testis sparing surgery is carried out in small tumours less than 2 cm, to avoid the sequelae of castration which include infertility, dependence on androgen replacement therapy.² Depending on the stage of the disease, histopathology and the serum markers, the adjuvant chemotherapy is given. Hormone replacement therapy in the form of testosterone is essential postoperatively. A thorough counselling of the patient regarding the risk of recurrence, infertility and surveillance is necessary. we can conclude-

- Cryptorchidism is the most common congenital genitourinary anomaly.
- The most common testicular tumour in cryptorchidism is seminoma testis.
- In a patient with abdominal mass and undescended testis, malignancy should be suspected.

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