Retroperitoneal Teratoma in an Adult With Carcinoid as the Main Component

Lt Col R Katoch^{*}, Brig Y Singh vsm⁺, Lt Col SPV Turlapati[#], Maj Gen Ramji Rai vsm^{**},
Lt Col V Srinivas⁺⁺

MJAFI 2002; 58 : 262-264

Key Words :Carcinoid; Retroperitoneum; Teratoma

Introduction

Teratoma are uncommon in adults. A case report in an adult is described. The histopathology revealed a teratoma with a main component of the tumour to be a Carcinoid. This combination has not been found in the literature till date. Most teratomas arise in infancy and are seen in the sacrococcygeal region [1,2]. They are also seen in the retroperitoneum, mediastinum and other parts of the body. The presentation in adults is uncommon. The frequency of malignancy increases with age [3]. The incidence of endocrine carcinoma in the retroperitoneum is extremely low [4]. Carcinoid tumours usually involve the gastrointestinal tract or the bronchial tree [5,6]. Retroperitoneum is a very rare site [6]. In searching literature we could not find any case of a retroperitoneal teratoma in an adult with Carcinoid as the main component.

Case Report

A 35 year old lady presented with a lower midline incisional hernia of 5 years duration and a lump in the abdomen of 4 years, with no complaints. Her bowel movements and menstrual periods were normal. She had undergone two Caesarian operations in 1989 and 1991. The general examination was normal. The abdomen showed an infraumbilical incisional hernia 15x8 cm, which was uncomplicated. A hypogastric firm non-mobile lump was palpable. On internal examination the lump was not palpable and the uterus was normal. Haemogram and biochemical investigations were normal. An ultrasound scan showed a solid mass just above the uterus and close to the iliac vessels, the liver and other structures were normal. Gynaecological examination was normal and chest radiograph did not show any abnormality. The patient was operated on 25 November, 1998, the abdomen was opened through the previous midline incision. A firm round mass was found just below the aortic bifurcation and flaying the iliac vessels laterally. The mass was dissected from the structures and removed after ligating the large feeder vessels (Fig 1). The uterus, ovaries, liver and the para-aortic lymph nodes were normal. The retroperitoneum was drained and the incisional hernia repaired.

Post operatively she did well and was discharged after ten days.

Pathological findings :

Gross : A 10x7x5 cm tumour with smooth bossillated external surface was seen. On cutting it revealed a well encapsulated light brown loosely granular tumour, showing a bony hard mass towards one pole. No cystic areas or dermal appendages like hair were seen (Fig-1).

Microscopic Examination : The tumour was encapsulated and predominantly made up of monomorphic polygonal to columnar cells arranged in solid islands, ribbons, festoons, and also at places in acinar configuration, separated by thin fibrovascular septa (Fig-2). The cell borders were indistinct. The nuclei were monomorphic, round to oval with "salt and pepper" chromatin. In addition there were other elements like cysts lined by stratified squamous epithelium, clusters of serous glands, mucous glands, mature bone, cartilage, smooth muscle bundles and glial tissue (Fig-3). No primitive neural elements or embryonal carcinoma like areas were seen. The tumour was immunohistochemically positive for chromogranin (Fig-4) and neuron specific enolase (NSE) (Fig-5) in carcinoid areas, but negative for synaptophysin. Electron microscopy revealed cytoplasmic dense core neurosecretory granules consistent with carcinoid.

Discussion

Teratomas primarily are congenital neoplasm with a wide spectrum of clinical characteristics. The neoplasm originates in pluripotent cells with a wide diversity of histological features foreign to the organ or anatomic site in which they arise. The most widely accepted theory is the germ cell theory. The primitive germ cells are totipotent and by the nineteenth day of intrauterine life are incorporated in the hindgut [7]. This explains why teratomas are common in the gonads, midline and paramedian locations. The extragonadal tumours are common in the hollow of sacrum generally along with spina bifida, mediastinum is the second commonest site and 70% of these occur in fe-

^{*}Reader in Surgery, ⁺⁺Reader in Pathology, Armed Forces Medical College, Pune - 411 040, ⁺Commandant, Military Hospital, Agra.
[#]Classified Specialist (Pathology), Military Hospital, Allahabad, ^{**}Commandant, Army Hospital (R&R), Delhi Cantt-110 010.

Fig. 1 : Photograph of the tumour from the retroperitoneum and cut open

Fig. 2 : Carcinoid tumour showing the cellular arrangement in ribbons and festoon magnification x 20

Fig. 3 : Microphotograph showing a cyst lined by stratified squamous epithelium, island of bone and carcinoid tumour (10x)

males [7]. These may arise in retroperitoneum rarely. The occurrence of teratomas is 50% by first year and 75% by 5 years of age [7]. Teratomas are usually ben-

Fig. 4 : Photomicrograph showing focal positivity for chromogranin by immunohistochemistry (40x)

Fig. 5 : Photomicrograph showing diffuse positivity for NSE by immunohistochemistry (20x)

gin and consist of fully differentiated mature tissue, and with advancing age the proportion of malignant teratomas increases [3]. Tezel et al reported 25% teratomas in a literature search of retroperitoneum teratoma [8]. Neonatal teratomas may recur in adulthood and give rise to malignancy [9]. The percentage distribution of benign, immature and malignant types, varies from 55-75%, 11.8-27.5% and 7.5-13.2% respectively [1,2]. The immature component is usually neuroepithelial with foci reminiscent of neuroblastoma and/or neuroepithelial tubule. Several case reports of extra-renal Wilm's tumour arising in sacrococcygeal and retroperitoneal teratomas have been recorded [2]. The malignant forms are the endodermal sinus (yolk sac) tumour and embryonal carcinoma [10]. The earlier names were carcinoma, teratocarcinoma, adenocarcinoma or papillary carcinoma. The occurrence of squamous cell carcinoma and mucinous adenocarcinoma in a pre-sacral teratoma has been described [8]. There have been very few reports of endocrine carci-

noma in the retroperitoneum, whether functional or not [4]. The speculation is that the origin of such tumours is likely to be from ectopic group of cells misplaced during embryonic development. It could be a monomorphous-monodermal form of teratoma or the gut itself becoming pedunculated and then marsupialised to the ventral space [4]. Retroperitoneal teratoma may present as an acute abdomen in elderly persons. Carcinoid tumour is classified as neuroendocrine tumour and shares cytochemical features with other endocrine tumours. Most originate in the bronchus, appendix, rectum and jejunioileum [5]. The rate of metastasis of small intestine Carcinoid is only 20-30% [5]. Carcinoid may remain asymptomatic or manifest with carcinoid syndrome, local invasion and mass effect or metastasis. Carcinoid of the kidney has been described. Carcinoid of the retroperitoneum is very rare and only few cases have been recorded earlier, one as a case of massive retroperitoneal bleeding [6] and another of retroperitoneal ureteric obstruction. A case of primary testicular carcinoid tumour with teratoma has been described. In this case report, a retroperitoneal teratoma was found in an adult woman with a Carcinoid component, which was asymptomatic, and this combination has not been recorded. The patient has been followed for one year with no metastasis or carcinoid syndrome, and tumour markers AFP and beta HCG have been normal.

References

1. Mahour GH, Wooley MM, Trivedi SN et al. Teratomas in infancy and childhood. Experience with 81 cases. *Surgery* 1974;76:309-18.
2. Valdiserm RO, Yunis EJ. Sacrococcygeal teratoma. A review of 68 cases. *Cancer* 1981;148:217-21.
3. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratomas. American Academy of Paediatric Surgical section survey; 1973. *J Paediatric Surg* 1974;9:384-42.
4. Yajima A, Toki T, Morinaga S et al. A retroperitoneal endocrine carcinoma. *Cancer* 1984;54:2040-2.
5. Godwin JD. Carcinoid tumours : An analysis of 2837 cases. *Cancer* 1975;36:560-9.
6. Rungta V, Vaiphei K, Goenka MK et al. Carcinoid tumour presenting as acute abdomen due to haemoperitoneum. *Indian J Gastroenterol* 1995;14(4):147-8.
7. Tapper D, Sawin R. Teratomas and other germ cell tumours. In : O'Neill JB, Rowe MI, Grosfeld JL, et al, editors. *Paediatric Surg*, St Louis, Missouri, Mosby, 1998;407-54.
8. Tezel E, Sare M, Edali N et al. Retroperitoneal malignant teratoma; A case report. *Mater Med Pol (Poland)* 1995;27(3): 123-5.
9. Lahdenne P, Heikinheim M, Nikkanen V et al. Neonatal benign sacrococcygeal teratoma may recur in adulthood and give rise to malignancy. *Cancer* 1993;72:3727-31.
10. Ablin A, Isaac H Jr. Germ cell tumours. In : Pizzo PA, Poplack DG, editors : *Principles and Practice of Paediatric Oncology*; Philadelphia, JB Lippincott, 1989:713-31.