IDIOPATHIC RETROPERITONEAL FIBROSIS

(Review of Literature and Report of Three Cases)

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The term 'Idiopathic Retroperitoneal Fibrosis' (I.R.F.) was first coined by Ormond (1948) for the cases presenting with ureteric obstruction due to mass of fibrous tissue of unknown etiogenesis. Since then, sporadic reports have appeared in world literature (Ormond, 1960; Mitchinson, 1965). The entity is peculiar to gynaecology as most of the presenting symptoms including low backache, lower abdominal pain, costo-vertebral or lumbar pain, tender flanks, frequency of urine or oligouria are related to genito-urinary tract, but invasion of uretric wall is strikingly absent.

Ormord (1960) reviewed the literature critically and classified the cases into 2 distinct groups:

(A) Where fibrosis involves both ureters coincidentally or in a succession and there is no pre-existing or co-existing cause of inflammation. These are true cases of I.F.R.

(B) Where the fibrosis is more localized, and there is causal connection with pre-existing or co-existing inflammation as in cases of endometriosis or past irradiation and are mistaken for I.R.F.

Present study of 3 cases, chiefly presenting with gynaecological symptoms, is the first available report from India. The aim of the study is to pin-point the etiological factor/factors.

CASE REPORT

Case 1

A poorly built 25 years old female was admitted with persistent dull lower abdominal pain for 4 months and frequency of micturation for 1 month. On local examination, there was a non-tender, soft, lobulated lump extending above the pubic symphysis upto 5 cms below the umbilicus in the midline. The margins of lump were ill-defined, it did not move with respiration, appeared to extend into the pelvis. On bimanual vaginal palpation uterus was anteverted, firm, normal in size and fornices were free. The lump was not related to uterus or adenexa.

Routine investigations, and plain X-ray abdomen did not reveal any positive findings except faint soft tissue shadow. The liver function

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tests revealed an altered A:G ratio. I.V.P. was not done. A clinical diagnosis of retroperitoneal tumour was made.

On laparotomy a non-encapsulated, soft, fleshy retroperitoneal mass was localised in the pelvis. It was not adherent to intestines but pelvic part of right ureter was embedded in it. The intraoperative frozen section revealed "bundles of fibrous tissue with occasional collection of inflammatory cells". There was no evidence of malignancy. An attempt was made to resect the mass. but a complete resection was not possible due to adherence with the common iliac vessels. The ureter was free. The patient was given routine postoperative care, the postoperative period was uneventful.

Case 2

A 35 years old female, gravida 4, was admitted with the complaints of low backache and chronic lower abdominal pain for 6 months and a palpable mass in lower left flank for 3 months. The pain was dull, unrelated to menstruation, urination. defaecation but relieved by antispasmodic and analgesics. She had normal menstrual cycles.

On examination, there was an on ill-defined lump in the left iliac fossa, lower margin was not definable as it extended into pelvis, non-tender, non-mobile, dull on percussion. On vaginal examination, uterus was anteverted and of normal size. adenexa free and lump had no relation with the uterus or adenexa.

Routine investigations and plain X-ray abdomen did not reveal any relevant findings except lowered serum protein with an altered A:G ratio. A clinical diagnosis of retroperitoneal tumour was made.

On laparotomy the lump was firm, non-capsulated but not infiltrating the adjacent viscera; iliac vessels and ureters were free. Whole of the mass was resected out. Postoperative period was uneventful and patient was discharged on 14th postoperative day in satisfactory condition.

Case 3

Patient aged 38 years, gravida 7, was admitted with the complaints of irregular vaginal bleeding for last 1 year, low backache. dull pain in the lower abdomen, not related to menstruation, urination and defaecation. On systemic and abdominal examinations there was no positive findings. On vaginal examination, uterus was enlarged to 8 weeks size, irregular. fornices were clear. On investigation lowered serum protein with altered A:G ratio was found. No other relevant positive findings. Diagnosis of fibroid uterus was made and total hysterectomy was planned.

On laparotomy, the uterus was enlarged, irregular; tubes and ovaries were normal. In addition, there was a diffuse, non-capsulated lump, arising deep from pelvis and retroperitoneal space. The lump was firm in consistency with irregular margins. After total hysterectomy, attempt was made to resect the mass which could not be taken out completely as both the ureters in their pelvic portion were deeply embedded. Both iliac vessels were free.

Pathology

Macroscopic: Surgical biopsy specimen in all the 3 cases were irregular, greyish white, soft to firm and fleshy pieces of tissues measuring 2.5 x 2 x 2 cms to 7 x 4 x 3.5 cms (6 pieces in Case 1. 5 pieces in Case 2 and 8 pieces in Case 3, weighing 250 gms, 150 gms and 400 gms each respectively). The cut surface was homogenous with few areas of whorling. Haemorrhage, necrosis and calcification were strikingly absent.

Microscopic: Histomorphological features were similar in all the 3 cases and revealed 2 patterns: (a) a less cellular area with more mature and immature collagen bundles which were arranged in irregular bundles. In between them the blood vessels were showing fibrinoid necrosis in the tunica media and their lumen was narrowed. Some of the blood vessels showed even complete obliteration of lumen. There was only mild mononuclear cell infiltration around the blood vessels and predominent cell was plasma cell. Polymorphonuclear cell infiltration was absent (Fig. 1).

(b) A more cellular area with proliferation of young fibroblasts with focal collection of mononuclear cell infiltration with even formation of microfollicles. However acute inflammatory changes were not seen. In between fibrous bundles, focal collection of lipid cells was also seen (Fig. 2).

Metachromatic stain was negative but Periodic-Acid-Schiff reaction and Alcian blue reaction were faintly positive in fibrinoid area. In rest of the tissue all were negative.

Etiopathogenesis

Various aetiological factors have been

put forward to explain its exact underlying nature but none of them make a clear connotation of the disease. These factors range from:

(i) Leakage of urine from forniceal rupture forming "Perinephric urine granuloma (Himman, 1960)", but it does not explain occurrence of lesion at other sites specially its relation to aorta and larger vessels.

(ii) Leakage of blood from aorta. Leaking aortic aneurysms may cause fibrosis (Heckett, 1958), but lack of haemosiderin in extracellular space or in macrophages rules out this possibility. Moreover, the type of cellular infiltration favours the allergic reaction rather than reactive fibrosis.

(iii) Lymphatic obstruction: In some cases the inflammation specially of lymphatics around the aorta may lead to fibrosis in the adjacent area (Mathison and Holta, 1966) but upward flow of lymphatics and normal lymphangiograms in some cases does not correlate with the cause.

(iv) Infection: Urinary tract infection may cause fibrosis by spreading via gonadal veins or lymphatics (Shaheen and Johnston, 1959). In few necropsy studies, presence of normal lymphatics and lymphnode lateral to or above the fibrosis rules out this possibility. Secondly, histological and bacteriological examinations have failed to demonstrate etiological agents or any evidence of acute inflammatory response.

(v) Adipose tissue infection: Adipose tissue infection specially in Weber-Christian disease may be responsible for fibrosis (Mitchinson, 1965) but adipose necrosis is not a constant feature of this disease and is hence ruled out as a sole etiological factor.

(iv) Toxoplasma infection: Mitchinson (1969) pointed out that there is a slight increase in antibody titre against toxoplasma but histologically there was no evidence of the parasite. This increased serological titre may be incidental and does not hold good in every case.

(vii) Connective tissue disease (autoimmune disease). Pugh (1960) pointed out that this is a autoimmune disease similar to rheumatoid disease and pointed out the followng facts in its favours:

(a) Fibrinoid necrosis in the wall of blood vessels.

(b) Mononuclear cell (predominantly plasma cell) infiltration around the blood vessels.

(c) Absence of acute inflammatory cell infiltration.

(d) Avascular area with mature and immature collagen bundles.

(e) Absence of demonstrable microbial agent.

(f) Good response to corticosteroids.

In the present study, histological and clinical findings also favour the above view.

A short term follow up of 15, 12, 7 months did not reveal any recurrence of symptoms, favours a good clinical response even with surgery alone.

Summary

Idiopathic retroperitoneal fibrosis (I.R.F.) usually presents with vague gynaecological symptoms, strikingly no relation with genito-urinary system. Various etiological symptoms have been

suspected but the findings favour a possible autoimmune genesis. Clinically the condition may be mistaken with retroperitoneal tumours. It gives a good response with corticosteroid therapy.

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See Figs. on Art Paper VII