RETROPERITONEAL FIBROSIS. PRESENTATION WITH NEOVASCULARITY ON ANGIOGRAPHY.

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SUMMARY
An unusual case of retroperitoneal fibrosis presenting an arteriogram with a pattern of neovascularity similar to that shown on malignant neoplasms is reported. Such a finding had hitherto not been described. The unusual observation of tumor neovascularity prompts the report of this proven case of retroperitoneal fibrosis.

RESUMO
Fibrose retroperitoneal. Imagem angiográfica de neovascularização.
Apresenta-se um caso de fibrose retroperitoneal, cuja arteriografia pélvica revelou vasos tumorais semelhantes aos das neoplasias malignas. Tal achado não tinha previamente sido descrito na literatura médica.

CASE REPORT
This 63 year old white female was admitted owing to progressive swelling of the left lower extremity for the past month. The patient also complained of occasional nocturia and alternating diarrhea and constipation, but denied any history of chills or fever, anorexia, or recent weight loss.

Pertinent past medical history included surgery for gall-bladder, herniorrhaphy, and medical treatment for osteoarthritis. As treatment for her osteoarthritis, the patient had been medicated with Indocid 50 mg B.I.D. for the past five years.

Pertinent findings on physical examination were limited to a firm, 4-5 cm in diameter, tender mass localized in the left lower abdominal quadrant.

Pitting edema of the left lower extremity was noted.

The cardiovascular system was within normal limits. The blood pressure was 124/72.

Laboratory exams on admission revealed a hemoglobin of 8.9 g., a hematocrit of 28%, WBC - 7,200, BUN - 38 mg%, creatinine - 2.4 mg%. Urinalysis revealed 100 WBC per large power field and its subsequent culture demonstrated more than 100,000 colonies of E. coli per cc.

RADIOGRAPHIC FINDINGS
A computerized radionuclide urogram suggested obstructed uropathy resulting in damage of both right and left kidney.

The intravenous urogram showed a delayed nephrographic stain on the left side. A 6 hour delayed film finally showed faint visualization of the pyelocaliceal system of the left kidney and evidence of moderate pyelocalycectasis and ureterectasis of the upper third of the left lateral wall of the bladder which was thought to be the cause for the obstruction of the left ureter (Fig. 1).

A left retrograde ureterogram demonstrated dilatation of the pyelocaliceal system in the upper ureter to the point of crossing at the pelvic brim. From this point distal the ureter appeared to be encased. (Fig. 2).

Figure 1: Intravenous urography: faint visualization of the pyelocaliceal system of the left kidney and deformity of the left lateral wall of the bladder, which was thought to be the cause for the obstruction of the left ureter.
With the provisory diagnosis of a neoplastic lesion involving the left lateral wall and extending to the lateral pelvic wall, most likely a carcinoma of the cervix, the patient was explored. A fibrous rock-hard retroperitoneal mass extending from the sacral promontory downward and filling the entire true pelvis was encountered. The gross appearance favored an extensive carcinoma of the rectum. However, frozen sections established the diagnosis of retroperitoneal fibrosis.

**DISCUSSION**

Retroperitoneal fibrosis was first described by Alberen in 1905. To date, more than 200 cases of retroperitoneal fibrosis have been reported.\(^1\)\(^,\)\(^2\)\(^,\)\(^3\) Nonspecificity of the clinical presentation has curtailed successful preoperative diagnosis and in most instances the diagnosis is made by surgical exploration and histopathologic examination of retroperitoneal tissues.

The intravenous urogram, in conjunction with the barium enema has provided the most specific criteria suggesting the diagnosis of retroperitoneal fibrosis. Encasement of the ureter in sheets of fibrotic tissue occasions a characteristic cessation of normal peristalsis\(^4\) which ultimately results in hydronephrosis of the afflicted renal unit. The abundant deposition of collagenous material in the retroperitoneal sheets is also responsible for the often observed characteristic medial displacement of the ureters at the level of the fourth and fifth lumbar vertebrae. Similarly, deposition of this abnormal tissue will result in encasement of the rectosigmoid with disturbance of function of this bowel segment.

In the past, preoperative diagnosis of retroperitoneal collagenosis has rested on characteristic changes afflicting the inferior vena cava and resultant drainage abnormalities in retroperitoneal veins. Encasement or occlusion of the inferior vena cava with resultant collateral flow through the hemiazygos, azygos, and lumbar veins has been advocated to substantiate the suspected diagnosis of retroperitoneal fibrosis.\(^1\)

Isolated reports in the literature on arteriographic manifestations\(^6\)\(^,\)\(^7\) have emphasized encasement of aorta and iliac arteries. Apart the case of retroperitoneal fibrosis reported by Keys\(^7\) with hypervascularity, neovascularity was not previously reported in medical literature.
Since deformity of the bladder secondary to deposition of fibrolipomatous tissue can mimic the changes resultant from the primary bladder neoplasm or a neoplasm secondarily involving the bladder, such as carcinoma of the cervix, arteriography might well be called upon to differentiate fibrolipomatosis and retroperitoneal collagenosis from a malignant neoplasm on basis of documentable tumor neovascularity. The observations in our patient showing an identical arteriographic pattern to that theme with tumor neovascularity of primary or secondary neoplasms involving the bladder eliminates arteriography as an effective modality for differentiation of these entities and appears to endorse definitive diagnosis upon thin needle biopsy.

REFERENCES


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Our patient demonstrated encasement and neovascularity usually considered characteristic of that found with malignant neoplasms. The fibroblastic proliferation with pleomorphic inflammatory reaction within the partial compartments 1 may be the anatomic basis for the neovascularity.