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EDITORIAL

PERIURETERITIS FIBROSA

In the crop of clinical entities that each year produces, none is likely to be more perplexing than periureteritis fibrosa—probably the most recently recognized distinct disease entity arising in the retroperitoneal space. The most concrete definition of the condition is to describe its pathology: a non-specific progressive fibrosis in the peri-ureteric fascial space, which eventually envelops the ureters and disturbs their function. Beyond that statement of fact, all is vague. The clinical picture is bizarre in the extreme, presenting in a variety of ways—backache, abdominal pain, sudden anuria or slow impairment of renal function. Laboratory and X-ray tests may be negative in the beginning, and thereby allay the doctor's fears for the health of his patient; or, worse still, the persistence of symptoms in the absence of signs may drive him to diagnose a functional disorder.

The great difficulty, then, is recognition. Pathological conditions of the retroperitoneal space are notoriously difficult of diagnosis. One arresting symptom, said to be helpful when it occurs, is the peculiar posture adopted by the patient with severe pain from the condition; he prefers to lie with his face down, 'buckling over the edge of the bed'[1]—perhaps to obtain the relief which gravity can afford him. Another diagnostically important feature is the obvious presence of pain in the absence of physical signs. This is said to be characteristic of an expanding retroperitoneal lesion, but it would be easy to fob off the patient—particularly if the ancillary diagnostic tests are negative or inconclusive, as they frequently are—with a lotion for his backache, an antacid for his ulcer, or a diuretic for his kidneys. There are no signs of specific inflammation—the erythrocyte sedimentation rate remains normal, pyrexia is late and due to secondary complications, no regional lymphadenitis occurs, and all biopsy material has been consistently negative on laboratory culture and inoculation for tuberculosis. Moreover, the condition appears to be self-limiting; patency of the urinary outflow from the kidneys may become re-established without apparent cause. Radiography may do more harm than good, for concomitant *bona fide* lesions may be discovered which have the effect of diverting attention from the true diagnosis. In one of Raper's cases an unrelated peptic ulcer was found;[2] in another case (complaining of backache) a malignant spinal tumour was suspected and the patient submitted to deep X-ray therapy before the true diagnosis of periureteritis was made.[3] Intravenous pyelogram may show a unilateral unexplained

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Van al die siektes wat elke jaar in die kliniek as eenhede uitgeken word, is periureteritis fibrosa—die jongste selfstandige siekte-eenheid wat agter in the buikholte ontstaan—seker die verwarrendste. Die mees konkrete definisie van hierdie siektetoestand is die beskrywing van sy patologie: 'n onbepaalde, toenemende fibrose van die bindweefselspasie rondom die urinebuis, wat uiteindelik die ureters omgewe en hul werking belemmer. Afgesien van hierdie feiteverklaring is alles vaag. Die kliniese beeld is uiters bizar; die siekte kan op verskillende wyses presenteer—rugpyn, buikpyn, skielike urienloosheid of geleidelike versteuring van nierwerking. Aan die begin kan die laboratorium- en X-straaltoetse negatief wees en dus die dokter gerus stel oor sy pasiënt se toestand; of, nog erger, die voortduur van simptome terwyl siektetekens uitbly kan hom dwing om 'n funksionele stoornis te diagnoseer.

Die groot moeilikheid is dus om die siekte uit te ken. Patologiese toestande van die retroperitonele holte is berug om die moeilikhede wat hulle by diagnose oplewer. 'n Merkwaardige simptoom, wat glo baie behulpsaam is wanneer dit voorkom, is die eienaardige houding wat die pasiënt ineem as hy baie pyn weens dié siekte verduur: hy lê liefs op sy maag, omgekrul oor die rand van die bed[1]'—miskien omdat hy op dié manier weens swaartekrag verligting vind. Nog 'n diagnostiese kenmerk is dat die pasiënt duidelik pyn verduur hoewel daar liggaamlik geen tekens gevind kan word nie. Na bewering is dit tipies van 'n groeiende retroperitonele letsel maar, veral as die bykomende toetse negatief of onoortuigend is, soos dit so dikwels gebeur, is dit maklik om die pasiënt te ‚paai' met 'n smeermiddel vir sy rugpyn, 'n teensuur vir sy maagseer of 'n diuretiese middel vir sy niere. Daar is geen tekens van spesifieke inflammasie nie—die eritrosiet-besinkingspoed bly normaal, koors verskyn eers later en dan is dit die gevolg van sekondêre komplikasies, daar is geen streek-limfklierontsteking nie, en alle biopsiemateriaal was nog altyd negatief by laboratoriumkweking en inenting vir tering. Ook blyk dit dat die toestand sigself beperk: die urienuitskeiding uit die niere kan ewe skielik sonder enige ooglopende rede weer vryelik begin. Radiografie kan meer kwaad as goed verrig omdat ander *bona fide* letsels óntdek kan word wat die dokter kan mislei en die eintlike siekte kan maskeer. By een van Raper se gevalle is 'n onverwante maagseer ontdek;[2] by 'n ander pasiënt (wat oor rugpyn gekla het) was 'n kwaadaardige ruggraatgewas vermoed en is die pasiënt onderwerp aan diep X-straalterapie voordat die eintlike kwaal van periureteritis gediagnoseer is.[3] 'n Binneaarse piëlogram kan moontlik 'n eensydige, onverklaarde hidronefrose aandui, of miskien slegs oopgesperde kelke met 'n donker uretersegment

hydronephrosis, or perhaps merely dilated calyces with an obliterated ureteric segment—which the radiologist may be excused for assuming to be due to spasm from ureteric peristalsis. In the cases so far described (mostly from the Massachusetts General Hospital[3, 4]) ureteric involvement with consequent renal functional impairment was a late feature of the condition. One recent case underwent laparotomy after 4 months of vague abdominal pain and weight loss but no renal impairment. At operation an ill-defined retroperitoneal mass was found in the lumbar region, which was considered by the surgeon to contain necrotic areas but which upon microscopic section showed merely 'subacute and chronic inflammation with fibrosis'. When the surgeon released the ureter from its fibrous web, the symptoms receded and the patient recovered. A second patient, presenting with acute anuria following several months of abdominal 'soreness', was found on laparotomy to have 'a retroperitoneal inflammatory process' that enveloped the ureters and caused obstruction to the outflow of urine. The ureters were freed and the patient recovered. It thus seems that this entity (for such it is) can sometimes only be diagnosed at operation—and then not invariably, unless it is specifically suspected or biopsy material taken. Certainly it represents a challenge to the skill of the whole clinical team.

1. Editorial (1957): Lancet, **2**, 780.
2. Raper, F. P. (1955): Proc. Roy. Soc. Med., **48**, 736.
3. Case Records Massachusetts General Hospital (1956): New Engl. J. Med., **255**, 90.
4. *Idem* (1957): *Ibid.*, **256**, 1198.

—en die radioloog kan vergewe word as hy aanneem dat dit die gevolg van kramp weens ureterperistalsis is. By die gevalle wat dusver beskryf is (die meeste is afkomstig uit die Massachusetts Algemene Hospitaal[3, 4]) was ureter-aantasting en gevolglike nierfunksiebelemmering 'n laatverskynsel by hierdie siekte. 'n Onlangse geval het na 4 maande van vae buikpyn en gewigafname, sonder nierbelemmering, 'n laparotomie ondergaan. Tydens die operasie is 'n vaag omskrewe retroperitonele massa in die lendestreek gevind, en die chirurg het gemeen dat dit nekrotiese areas bevat. By mikroskopiese seksie is dit egter bevind dat hierdie massa slegs ‚subakute en kroniese inflammasie met fibrose' was. Nadat die chirurg die ureter van sy fibreuse netwerk bevry het, het die simptome vervaag en die pasiënt herstel. By 'n tweede pasiënt, wat met akute anurie volgend op maandelange ‚seerheid' in die buik gepresenteer het, is dit by laparotomie bevind dat daar 'n ‚inflammasie-streek in die agter-buikholte' was wat die ureters omvou het en die vloei van die urien belemmer het. Die ureters is losgemaak en die pasiënt het herstel. Dit blyk dus dat hierdie siekte-eenheid (want dit *is* 'n siekte-eenheid) soms alleen operatief gediagnoseer kan word—en dan ook nie altyd nie, tensy dit spesifiek vermoed word of tensy biopsiemateriaal geneem word. Dit stel gewis die vaardigheid van die hele kliniese span op die proef.

1. Van die Redaksie (1957): Lancet, **2**, 780.
2. Raper, F. P. (1955): Proc. Roy. Soc. Med., **48**, 736.
3. *Case Records Massachusetts General Hospital* (1956): New Engl. J. Med., **255**, 90.
4. *Idem* (1957): *Ibid.*, **256**, 1198.

THE SPRUE SYNDROME

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Malabsorption from the gastro-intestinal tract is usually described by its most prominent feature, steatorrhoea. The presence of excessive amounts of fat in the stools leads to the passage of bulky, offensive motions. This is not always obvious, and the patient may present with secondary effects due to the deficient absorption of vitamins, minerals and calories, etc. In some cases the steatorrhoea is due to lack of digestive secretions, as in chronic obstructive jaundice and pancreatic disease; inefficient mixing of food with enzymes is thought to account for a proportion of cases of post-gastrectomy steatorrhoea.[54] It is not proposed to discuss these further, but to concentrate on the enterogenous steatorrhoeas, where the fault lies in the lumen or wall of the small bowel.

The enterogenous steatorrhoeas have many features in common (see below). There are 3 sub-groups: those cases with gross irreversible changes in the bowel wall, those where abnormal anatomical factors produce stagnation, and the sprue syndrome. Among the group with irreversible pathological changes are regional enteritis, lymphoma, Whipple's disease, amyloid, and scleroderma. Active tuberculous enteritis is probably a less important cause than it was thought to be, and Franz J. Ingelfinger[11] stresses that steatorrhoea is rarely due to lymphatic blockage by tuberculous mesenteric adenitis.

In the group of anatomical disorders stagnation of intestinal contents facilitates the growth of an abnormal bacterial flora with an avidity for essential nutritional factors.[24, 57] The ability of certain strains of *Str. faecalis*, when provided with bicarbonate and folic acid, to synthesize significant amounts of fats *in vitro* is of great significance in this regard.[48] The sufferer from the stagnation syndrome is thus deprived of important dietary constituents and, in addition, the remainder of the small bowel is irritated by overflowing exudates. The causes are multiple intestinal strictures,[9] blind loops of small intestine[14, 33] and jejunal diverticulosis.[2] In some, but not all, cases, strictures are due to healing of tuberculous girdle ulcers. Examples of blind loops are entero-anastomosis, gastro-jejuno-colic fistula, inadvertent gastro-ileostomy and internal fistula. Multiple jejunal diverticula are of congenital origin. Massive intestinal resection is not an important factor in producing a sprue-like picture if the remaining bowel is healthy.[33]

The remaining cases of enterogenous steatorrhoea may be considered as belonging to the 'sprue syndrome'. Admittedly such a definition, which depends on exclusion rather than on positive features, is hardly satisfactory, but prevailing knowledge of the syndrome has not permitted a more precise statement. However, newer discoveries (see below) indicate