Isolated Epididymal Vasculitis

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Scrotal mass may be caused by conditions such as local infection, or by an inflammatory or neoplastic process. It is rarely a manifestation of systemic disease such as polyarteritis nodosa. The treatment of each condition is specific and the incorrect diagnosis may lead to unnecessary orchiectomy or potential toxic immunosuppressive therapy. In this report we describe a rare case of isolated vasculitis of the epididymis presenting as a scrotal mass in a pubertal boy. We also review the diagnostic possibilities including isolated PAN.

Case Description

A 13.5 year-old boy was referred to our surgical department with left scrotal pain that had begun 7 days earlier. On examination, the epididymis of the left testis was thickened and tender and a sensitive localized swelling of about 1.5 cm in diameter was palpated at the lower pole of the testis. No skin or joint lesions were noted and the rest of the physical examination was normal. Urinalysis and cultures were negative. A full blood count, and liver and kidney functions were normal. The tumor markers alpha-fetoprotein, carcinoembryonic antigen, human chorionic gonadotropin and carbohydrate antigen 19-9 were not elevated. Ultrasound imaging of the testis showed a hypoechoic mass that was attached to the tail of the epididymis. A computed tomography scan of the abdomen, pelvis and lungs was normal.

On surgical exploration the epididymis was thickened, hyperemic and firm in consistency. The testis appeared normal. An open biopsy was taken from both the epididymis and the swelling that was attached to its tail. The frozen section from the specimen showed no sign of malignancy. Histological studies of the left testis subsequently demonstrated vasculitis of medium and small arteries. Also visible were regions of fibrinoid necrosis with thrombosis and perivascular fibrosis [Figure 1]. These findings were compatible with PAN. A comprehensive evaluation for systemic involvement was then conducted. Relevant laboratory findings revealed: erythrocyte sedimentation rate 50 in the first hour, C-reactive protein 18.3 mg/dl, and normal urinalysis, serum protein electrophoresis, immunoglobulins, and complement C3 and C4. Rheumatoid factor, antinuclear antibody, antineutrophil cytoplasmatic antibodies, hepatitis B surface antigen and antibodies to hepatitis C were negative. Chest X-ray, electrocardiogram, echocardiogram, and fundoscopy were normal.

Comment

Most cases of male reproductive system vasculitis (testis or epididymis) occur as part of a systemic disease, most commonly polyarteritis nodosa. It is seen less frequently in other vasculitides such as Wegener's granulomatosis, Henoch-Schonlein purpura, Goodpasture's syndrome, and rheumatoid arthritis, or due to infection such as typhus and Rocky Mountain spotted fever [1]. The process may be seen as an acute inflammation or as a discrete mass. Our patient had no fever, abdominal pain or arthralgia, and his physical examination revealed no skin lesions, arthritis or rheumatoid nodules. Laboratory tests showed normal urine and absence of serum autoantibodies, and chest X-ray was normal. This presentation of isolated vasculitis of the epididymis without other manifestations, together with the histological findings described, support the diagnosis of isolated PAN. Other diagnoses such as rheumatoid arthritis, Henoch-Schonlein purpura, Wegener's granulomatosis and Goodpasture's syndrome seem highly unlikely.

In systemic PAN, clinical vasculitis of the male reproductive system (testis or epididymis) occurs in up to 18% of patients [2]. Yet, autopsy has demonstrated an incidence of 60–86% involvement [2]. The affected testis and epididymis may be swollen with or without tenderness or pain, or they may appear completely normal. Other organs that have been reported to be involved are the appendix, gallbladder, breast, uterine corpus, and cervix [3]. These manifestations may be asymptomatic and the diagnosis can be made by histological examination. To the best of our knowledge, only one case of isolated PAN involving the male reproductive system has been previously reported in the pediatric age group. This was found incidentally in an undescended testis of a 14 year old boy who had undergone orchiectomy [3].

It appears that the majority of cases of isolated PAN in adults does not progress to the systemic form. Thus, it

PAN = polyarteritis nodosa

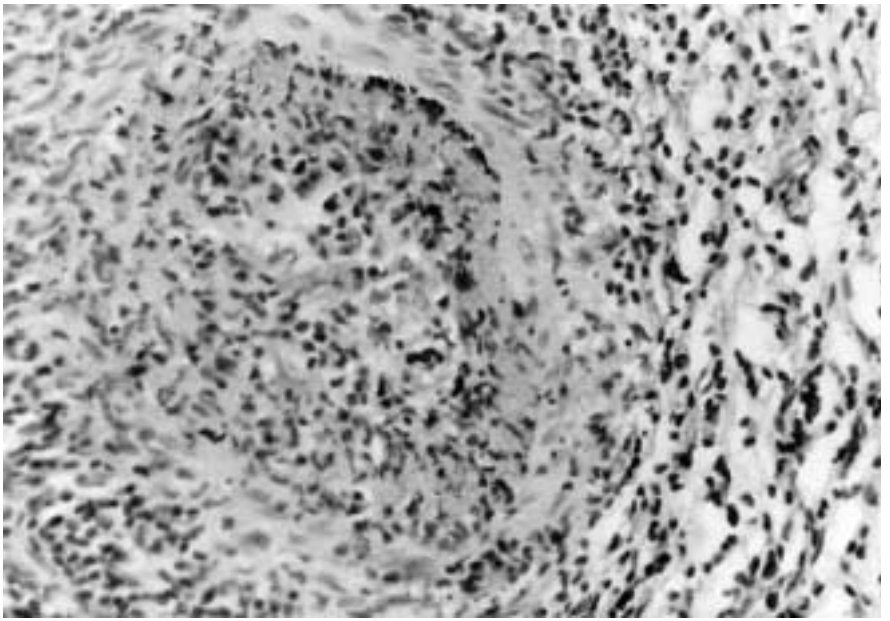

Figure 1. Epididymal biopsy demonstrating an artery with fibrinoid necrosis, inflammatory infiltrate of the wall, and thrombosis (hematoxylin and eosin).

is not surprising that the recommendations in the literature are for either a local excision or a short course of corticosteroids along with adequate clinical and serological monitoring for possible progression to systemic involvement [4]. In some reported cases, the testicular masses were diagnosed as tumors, and consequently the patient underwent a radical orchiectomy [3,5].

In view of the fact that the general condition of our patient was satisfactory and that his erythrocyte sedimentation rate and C-reactive protein level had returned to normal without any additional treatment, it was decided to follow the patient without initiating corticosteroid therapy. After 2.5 months the epididymal swelling had disappeared and the shape of the epididymis returned to normal. This was confirmed by ultrasound examination. He has now completed a follow-up period of 24 months and has no clinical or laboratory evidence of disease.

In this case we demonstrated that isolated PAN can resolve without corticosteroid therapy or total excision of the involved organ. However, we cannot exclude the possibility that the administration of a non-steroidal anti-inflammatory drug (diclofenac sodium) for 7 days following surgery – given because of the possibility of an inflammatory process – suppressed the inflammatory state. We conclude that in the pediatric age group, the possibility of isolated PAN must be considered when a testicular mass is found.

References

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