

Intramural ureteric calcification in dermatomyositis mimicking pelviureteric junction obstruction

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ABSTRACT

Juvenile dermatomyositis classically manifests as necrotising vasculitis in multiple organs. It mainly involves the vessels of skin and muscle and is associated with subcutaneous deposits of calcium. Ureteric involvement in dermatomyositis is extremely rare. The authors present a case of a 14-year-old girl with juvenile dermatomyositis with intramural ureteric calcification mimicking pelviureteric junction (PUJ) obstruction.

KEY WORDS: Dermatomyositis, ureteric calcification, pelviureteric junction obstruction

INTRODUCTION

Juvenile onset dermatomyositis is an idiopathic inflammatory myopathy that is first detected in childhood or adolescence. It is rare and affects 1.9 children per million in United Kingdom and Ireland.^[1] The estimated incidence in the United States ranged from 2.5 to 4.1 per million. The estimated annual incidence rates were 3.4 for white non-Hispanics, 3.3 for African American non-Hispanics and 2.7 for Hispanics. Girls are affected more than boys (2.3:1).^[2]

It is characterized by a rash accompanying or more often preceding muscle weakness. Calcification is the most characteristic soft tissue abnormality in dermatomyositis and is manifested by calcareous deposits in the intermuscular fascial planes or subcutaneous tissue. We report a rare case of dystrophic intramural ureteric calcification causing symptomatic obstruction.

CASE REPORT

A 14-year-old girl with juvenile dermatomyositis was referred with chronic right loin pain. On clinical examination she had characteristic cutaneous eruptions and muscle weakness. There was some fullness in the right loin. An ultrasound scan showed a right hydronephrosis, with no dilatation of the ureter. A MAG3 scan showed decreased function with poor drainage on the right. She was being treated with cyclophosphamide, methotrexate,

mesna, steroids and local applications. Due to the severity of her overall condition it was decided to manage the PUJ obstruction by the placement of a JJ stent. This relieved her loin pain. Repeat scans showed a reduction in hydronephrosis and improved renal drainage. Six months after the procedure she was readmitted for recurrent right loin pain. An X-ray showed calcification around the JJ stent. Replacement of the JJ stent proved impossible. Hence an emergency exploration was carried out. At operation there was narrowing of the ureter just below the PUJ. The pelviureteric junction was itself patent, but surrounded by a severe inflammatory reaction. The child underwent a dismembered pyeloplasty with excision of the upper ureteric segment. Post-operative recovery was uneventful. Histopathology of the upper ureter showed ulceration with marked fibrosis disrupting muscle coat, accompanied by abundant calcific debris in the lumen and sub epithelial stroma (Figure 1). Follow up scans 6 months after the procedure showed mild dilatation of the renal calyces with good drainage and function.

DISCUSSION

Juvenile dermatomyositis is a multi system disease characterised by necrotising vasculitis in multiple organs. It is probably the result of a humoral attack on the muscle capillaries and small arterioles. As the disease progresses, the capillaries are destroyed and the muscles undergo microinfarction. Dystrophic calcification occurs in 10-50% of affected patients.^[3-5]

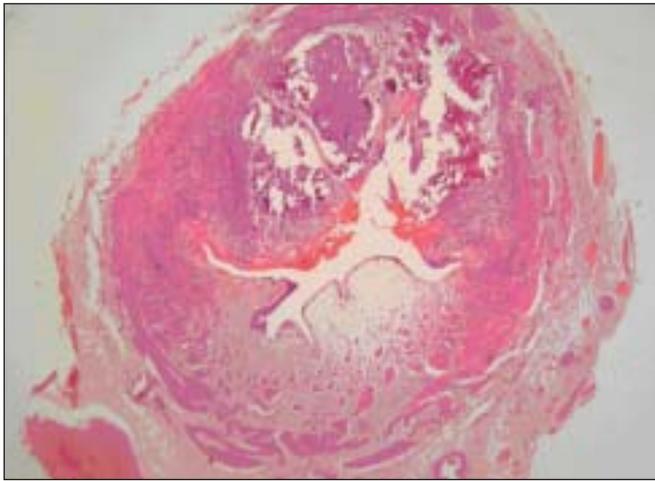


Figure 1: Low power section of the ureter showing intra mural calcification

In our patient, intramural ureteric calcification was the cause of the ureteric obstruction causing hydronephrosis and loin pain. Ureteric calcification has been described before.^[6,7] However, this has always been in the middle and lower thirds. Interestingly in our patient, the upper ureter was the area that was affected and calcification was noticeable on plain X-ray.

Dystrophic calcification in dermatomyositis can affect the

ureter. It can and does cause obstruction severe enough to affect the function of the relevant kidney. This can be managed temporarily by JJ stenting. Operative repair in the form of pyeloplasty or excision and uretero-ureteric anastomosis though difficult is successful in preserving renal function.

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