Efficacy of infliximab in a patient with refractory idiopathic retroperitoneal fibrosis

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Key words: infliximab, idiopathic retroperitoneal fibrosis

Case report

A 60-year-old woman presented in June 2006 with fatigue, abdominal pain, anorexia, and weight loss. She was receiving anti-hypertensive drugs for arterial hypertension. The erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were raised at 85 mm/1st h and 4.60 mg/dl (normal value <0.50 mg/dl), respectively. Autoimmune serology and tests for common infectious diseases including tuberculosis were negative. A chest radiograph was unremarkable, while tumour markers and fecal occult blood were within limits. Further imaging was requested to investigate the cause of the patient’s complaints.

Computerised tomography (CT) of the chest and abdomen showed concentric vessel wall thickening of the ascending thoracic aorta consistent with vasculitis, as well as a homogeneous perivascular cuff (maximum periaortic thickness: 59 mm) isodense to muscle surrounding the abdominal aorta and iliac arteries. The perivascular tissue had also encased the right ureter leading to right-sided hydronephrosis.

IRF was diagnosed. Secondary causes of retroperitoneal fibrosis including radiotherapy, surgery, intake of sclerosing drugs, infections, and tumours were excluded by the clinical history, laboratory tests, and by the imaging findings that showed no atypical features of the retroperitoneal mass such as tissue heterogeneity. Erdheim-Chester disease was excluded by the lack of perirenal retroperitoneal fibrosis and of typical manifestations such as bone pain. Prednisone (1 mg/kg/day) was commenced with marked clinical improvement and normalisation of inflammatory markers within a few weeks. A stent was also

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CASE REPORT

Infliximab in idiopathic retroperitoneal fibrosis / M.G. Catanoso et al.

inserted into the right ureter with rapid resolution of the right-sided hydronephrosis. However, attempts to taper the prednisone dose below 7.5 mg/day resulted in three clinical flares that required an increase in the prednisone dosage. Methotrexate (15 mg/weekly) was thus added as steroid-sparing agent. Despite the addition of methotrexate, in July 2007 while on prednisone 7.5 mg/day and methotrexate 15 mg/weekly, the patient had a new flare characterised by fatigue, abdominal pain, and diffuse aches and pains. ESR was 72 mm/1st h, and CRP 7 mg/dl. Fluorodeoxyglucose (FDG) positron emission/computerised tomography (PET/CT) was performed to assess disease activity and extent. PET/CT revealed grade 2 FDG uptake around the abdominal aorta spreading from the origin of the renal arteries to the aortic bifurcation (Fig. 1A). SUV ratio (abdominal aorta and surrounding retroperitoneal mass/liver) was 1.20. A repeat CT scan showed only a slight reduction in the size of the abdominal periaortic cuff (maximum periaortic thickness: 57 mm).

Because of persistently active disease despite conventional treatment, adjunctive therapy with infliximab 5 mg/kg at week 0, 2, 6 and 8-weekly thereafter was commenced. The patient gave informed consent before infliximab treatment, which was approved by the local ethics committee.

Four months later, the patient was asymptomatic, while the ESR and CRP decreased to 25 mm/1st h and 0.41 mg/dl, respectively. Prednisone was successfully tapered off by March 2009.

Discussion

Patients with IRF are usually managed with glucocorticoids, often combined with immunosuppressants, but some patients are refractory to conventional treatment. There is evidence that TNF-α inhibitors may be efficacious in patients with resistant large-vessel vasculitis (13). In this regard, a recent report also confirmed the efficacy of the anti-TNF-alpha monoclonal antibody- adalimumab in a patient with large vessel vasculitis associated to sarcodosis refractory to corticosteroids and methotrexate (14). Therefore, although there is no data on the efficacy of TNF-α inhibitors in IRF, we decided to treat our patient with the TNF-α blocker infliximab. Our results showed a full clinical and laboratory response to therapy paralleled by an improvement in imaging findings. More specifically, the retroperitoneal mass as assessed by CT shrank, albeit only modestly, after the onset of infliximab therapy. On the other hand, PET/CT demonstrated reduction in FDG uptake by the retroperitoneal mass. Such a residual retroperitoneal mass characterised by slight FDG uptake following therapy is consistent with metabolically inactive tissue. (15-17).

In addition, infliximab therapy allowed discontinuation of glucocorticoids and removal of the right ureter stent. These findings convergently point to efficacy of infliximab in this case of refractory IRF. In conclusion, this case suggests that infliximab may be a useful and safe therapeutic option for patients with IRF refractory to the traditional immunosuppressive treatment. Controlled studies are warranted to confirms our results.
CASE REPORT

Infliximab in idiopathic retroperitoneal fibrosis / M.G. Catano et al.

References