

**RETROPERITONEAL FIBROSIS**

It was with great interest that I read the article on retroperitoneal fibrosis by Bellamy and Tomich in your January 2014 issue of the *CJEM*.<sup>1</sup> This uncommon cause for abdominal pain should be considered in the differential diagnosis in older males who present to the emergency department with vague symptoms of back pain, pelvic pain, abdominal pain, gastrointestinal or genitourinary symptoms. Surprisingly, the article made no mention of IgG4-related disease, a newly recognized fibro-inflammatory condition that is the probable cause of most cases of retroperitoneal fibrosis.<sup>2</sup> This condition has been identified in virtually every organ system. This diagnosis ties together many diverse diseases that were previously thought to be separate idiopathic conditions:

orbital pseudotumour, retroperitoneal fibrosis, Riedel thyroiditis, Kuttner tumour, Mikulicz disease, interstitial lung disease, tubulointerstitial nephritis, autoimmune pancreatitis, periaortitis (leading to aneurysms), sclerosing mediastinitis and other diseases. While the etiology is unclear, it has many features of an autoimmune disease. In contrast to the female predominance in autoimmune diseases, the typical patient is a middle-aged to elderly male. IgG4-related disease tends to form tumefactive lesions and has frequently led to patients being thought to have malignancy. Patients with retroperitoneal fibrosis should be investigated for other organ system involvement. Corticosteroids have been effectively used to reduce fibro-inflammatory infiltrates, but it appears that rituximab

may be a more effective and longer-lasting treatment.<sup>3</sup>

Thank you for describing one of those rare but important causes of lower abdominal or back pain. Not all retroperitoneal masses are lymphoma!

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**REFERENCES**

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