A 58-year-old woman presented to the emergency department after a minor fall, complaining of lower abdominal pain. A large, tender mass arising from the pelvis was palpable, in association with abdominal wall bruising. The patient believed that she may have had a "lump" there for several years. She had had a total abdominal hysterectomy and bilateral salpingo-oophorectomy for benign fibroids three years previously. She denied any gastrointestinal (GI) or genitourinary symptoms. She had no night sweats or weight loss. Routine blood investigations were normal.

A CT scan of her abdomen and pelvis with oral and IV contrast contrast was performed to investigate what was thought to be a rectus sheath haematoma. This has revealed abnormal retroperitoneal lymphadenopathy in the mid and lower Abdomen (Fig. A). Most of her bladder was replaced by a 16 x 12 x 10 cm soft-tissue density mass (Fig. B). There was mild dilatation of both ureters with mild left hydronephrosis. Appearances suggested a malignant process, given the retroperitoneal adenopathy. Ultrasound-guided biopsy of a right-sided 13 x 8 mm solid inguinal lymph node was performed; histology of this was inconclusive.

Following this, a rigid cystoscopy confirmed a grossly abnormal bladder with a diffuse, thick mass and a non-friable, pale-looking mucosa which was intact. Multiple resection biopsies were taken. MRI of pelvis the next day showed almost complete absence of a bladder lumen. The bladder wall was diffusely thickened, measuring 10 cms in places (Fig. C). Pathologically-enlarged iliac and retroperitoneal nodes were seen. Abnormal stranding of perivesical fat suggested transmural malignant infiltration. The histology of her bladder biopsies showed dense, diffuse infiltration by small lymphocytes, covered in part by intact urothelium. Microscopy and immunohistochemistry supported the diagnosis of low grade, diffuse, B-cell Non Hodgkin's Lymphoma (NHL). A diagnosis of primary malignant lymphoma arising in the urinary bladder was made. She was referred on to the Haematology service for further investigation and treatment. Given that her International Prognostic Index is 2-3/5, six cycles of RCHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone) chemotherapy were planned. After completing her treatment, she will be re-staged with appropriate scans.

Comment

About 40% of NHL present in extra-nodal sites, with the skin and the GI tract being the most frequent sites. Primary bladder lymphoma is a rare disease, responsible for only 0.2% of extra-nodal lymphomas. The disease is typically seen in middle-aged and elderly females. The frequency of NHL in the urinary bladder is higher than Hodgkin’s lymphoma. Presentation can be variable with symptoms including haematuria, pain, dysuria, nocturia, frequency or recurrent UTIs. Bladder lymphomas tend to occur at the base and the trigone of the bladder and usually form a sessile mass with normal urothelium. Cystoscopy commonly shows a solid tumour without surface changes. The surrounding mucosa is typically intact but in a very large tumour the mucosa can be ulcerated. Upper tract dilatation can occur secondary to mass effect and ureteric obstruction. Histopathological studies are essential for the diagnosis. It is often difficult to differentiate lymphoma from sarcoma or carcinoma on frozen section biopsy. Histologically, primary bladder lymphoma is most commonly low-grade NHL of the B-cell type. About 20% of B-cell bladder lymphoma is high-grade. Reported management of primary bladder low-grade NHL is varied. Partial or total cystectomy, radiotherapy, chemotherapy, or combined therapies have all been described. Surgery may be inappropriate in these patients, given the high success rate with either radiotherapy or chemotherapy. Repeat cystoscopy is recommended for follow-up of all patients.

In summary, this case highlights the difficulties in diagnostic differentiation relating to large pelvic masses in middle age females. In the differential diagnosis of bladder tumours, malignant lymphoma of the bladder should be considered. Despite the fact that primary bladder lymphoma is a rare condition, we should be aware of the excellent complete remission rates and good prognosis.

Department of 1. Urology and 2. Radiology, Mercy University Hospital, Cork, Ireland.