

Case description: An 83-year-old lady presented to the surgical out-patient clinic with upper abdominal pain radiating to left side of her chest. She had recent history of significant weight loss, approximately 22kg. Abdominal examination was unremarkable.

Results and Conclusions: Haemoglobin and Liver Function Tests were normal. Gastroscopy showed one large 3cm ulcer with a shaggy base and rolled over margin in the first part of the duodenum (D1) which appeared malignant. Initial histology confirmed adenocarcinoma of uncertain origin. CT scan showed a 2.9x3.7cm partially cystic mass lesion in the head of the pancreas that was locally invading into the first part of the duodenum, with no evidence of metastases. Immunohistochemistry showed strong Ca19.9 positive, favouring primary pancreatic origin.

Take home message: Pancreatic cancer presenting as an ulcer in D1 is very rare. If a suspicious looking ulcer is found endoscopically at D1, it should be biopsied. A CT scan of the abdomen is also important in the work-up of such cases.

<http://dx.doi.org/10.1016/j.nhccr.2017.10.012>

Changing standards in the treatment of desmoid tumors?

A Trippel^{1,*}, R Cathomas², C Michelitsch¹, M Furrer¹

¹ Thoracic and Vascular Unit, Department of Surgery, Cantonal Hospital of Graubünden, Chur, Switzerland

² Department of Oncology, Cantonal Hospital of Graubünden, Chur, Switzerland

Objectives: Desmoid Tumors (DT) are rare locally aggressive and never metastasizing mesenchymal tumors. They usually grow slowly and often present on the trunk or extremity, on the abdominal wall or intra-abdominally. Radical surgical removal has been considered the treatment of choice for many years, although high recurrence rates ranging from 30% to 40% have been reported.

Methods: A 40-year old woman was referred to our department with a 4cmx2cmx3cm painless hard tumor on the lateral wall of the right thorax. A histological diagnosis of a DT was made after a biopsy specimen was obtained. After initial R0 surgical resection the patient developed a chronic pain syndrome. Another surgical R0 resection was performed after local recurrence was noted in an MRI fifteen months postoperative. 30 months after the first operation and 17 months after the second operation a second local recurrence was seen. A third operation was planned but the patient chose not to undergo surgery and was referred to the medical oncologist. A therapy with tamoxifen was started and supplemented with sulindac after two months since a tumor progression was shown in the first follow up MRI.

Results: The follow up under the tamoxifen/sulindac therapy showed disease stabilization after 12 months and a complete tumor regression after 1½ years. The tamoxifen/sulindac regimen and the follow-up were continued, and the patient is now tumor free after 3½ years of medical treatment. The pain syndrome has also improved although the patient is still under oxycodone therapy.

Conclusions: In current practice, the treatment of desmoid tumor medical therapy was reserved to unresectable disease or where radical resection would have brought to major functional or cosmetic losses. This present case calls into question the established surgical therapy of desmoid tumors. Although a strong evidence-based treatment is still missing, other recent studies have also suggested alternative different therapeutic strategies based on a “wait and see” strategy may be effective in the case of asymptomatic disease.

<http://dx.doi.org/10.1016/j.nhccr.2017.10.013>

A case report of metastatic renal oncocytoma

Gionathan Amante*, Yan Mei Goh, Henry Andrews

Milton Keynes University Hospital, Milton Keynes, Buckinghamshire, UK

Introduction: Renal oncocytomas are renal tumours that are largely considered to be benign. They represent 5.5% of all renal tumours and the age-standardised incidence is 0.3 per 100,000/year in men and women.¹ Despite this, there have been reported cases where these tumours have developed metastases. We report the case of a patient who had a right radical nephrectomy for renal oncocytoma who then presents nine years later with a pathological neck of femur (NOF) fracture secondary to metastases, suggesting malignant behaviour of the tumour.

Case description: A 62-year-old lady presented in December 2010 with shortness of breath on exertion, frontal headache and malignant hypertension. She was treated with Ramipril and her blood pressure subsided. On discharge she underwent further investigations, including 24hour urine catecholamine test – (normal), and abdominal ultrasound which revealed a mass in the right kidney. Computerized tomography (CT) confirmed a well-defined lower pole right renal solid mass measuring 7.5x5.7cm, extending to the renal pelvis with compression of the vascular pedicle. There was no evidence of extrarenal extension. Renal cell carcinoma stage T2 was suggested. She underwent a right laparoscopic nephrectomy and recovered well from the operation. Histology confirmed oncocytoma with atypical features of extrarenal invasion, absence of Hale's colloidal blue staining, negativity for cytokeratin 7 and positive vimentin staining. This patient had yearly follow-up CT. Four years after her first surgery, a nodule was noted in the right nephrectomy bed. Surgical excision confirmed recurrent oncocytoma. Two years after this she presented with pathological fracture of her right neck of femur. A bone biopsy confirmed metastatic disease. She had a dynamic hip screw fixation.

Results and Conclusions: The World Health Organization classifies renal oncocytoma as benign neoplasm.² Nevertheless there are case reports of metastatic disease in these tumours. Studies have revealed a classical pattern with typical features of benign neoplasia in 57.5–67% of cases. Malignant features such as atypical histological pattern, focal necrosis, multinucleation, mitotic activity, degenerative atypia and invasion of extrarenal fat have been described in others. Despite this, not all patients with such features progress to developing metastatic disease at 5-year follow-up. In fact only 2 patients out of 42 with atypical features developed metastatic disease.³ This suggests that there is a potential for metastatic transformation of this tumour, especially in the group with features of atypia. There is a however, paucity of information in published literature regarding this. The risk stratification of these atypical features will be a good start. Clearly, further research by way of genetic assessment, including chromosomal and tumour markers is required, as part of future diagnosis and management algorithm.

Take home message: Metastatic renal oncocytomas are extremely rare. Little is known about the atypical features that predispose metastatic transformation. Our patient developed local recurrence and a distant metastatic lesion with fracture neck of femur 6 years later. Our extensive search of literature has shown only 2 other reported cases of metastatic oncocytomas in the bone being the tibia and femur. The patient with the femoral fracture had multiple bone metastatic lesions. Further research into the biological features and characteristics of this extremely rare condition is required. This may help stratifying patients into metastatic risk potentials. Perhaps treatment plans can be tailored accordingly.

References

- 1) Gudbjartsson T, Hardarson S, Petursdottir V, Thoroddsen A, Magnusson J, Einarsson GV, Renal oncocytoma: a clinicopathological analysis of 45 consecutive cases. *BJU Int.* 2005 Dec;96(9):1275-9.