Symptomatic steroid induce multifocal diaphyseal bone infarcts treated with intra-medullary nailing

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Introduction: Non traumatic osteonecrosis is the ischaemic death of cellular elements within bone. Etiological factors implicated include long term corticosteroid use, alcoholism, sickle cell disease, systemic lupus erythematous, amongst others. Common sites of involvement include the proximal femur, knee, shoulder, ankle. Metaphyseal-diaphyseal lesions have been well described radiologically, however are commonly considered asymptomatic. There is thus a paucity of literature describing techniques used for symptomatic diaphyseal or metaphyseal lesions not involving the epiphyseal region.

Case description: Our patient is a 40-year-old woman diagnosed with Arnold-Chiari malformation in 2005 who was then surgically treated with foramen magnum decompression. In 2010 she was treated with 4 months of Dexamethasone 2mg for chemical meningitis. She presented to the Orthopaedic outpatient clinic in 2012, 16 months after ceasing steroid medication, with a 6 month history of difficulty walking due to pain in bilateral groins and bilaterally along her shins, left worse than right. MRI of both hips demonstrated anterior serpiginous lesions within the femoral heads consisted with AVN (Ficat II). MRI of lower legs showed isolated bone infarct in the metaphyseal-diaphyseal region of her tibias bilaterally. She had bilateral total hip arthroplasties with immediate relief of hip symptoms. Our patient underwent bilateral tibial intramedullary nailing using a Stryker T2 nail with a medial parapatellar approach. At both the 6 week and 5 month follow-up she had no further pain, was non tender to palpation and was very satisfied with result.

Results and Conclusions: We are unaware of any reports of the development of symptomatic diaphyseal osteonecrosis in patients receiving corticosteroids for the treatment of meningitis. Much of the literature regarding management of osteonecrosis is focused on the treatment of epiphyseal lesions in the femoral head and around the knee. Diaphyseal lesions have been well described radiologically but are often defined as asymptomatic and clinically insignificant. Our use of intramedullary nailing thus illustrates an effective surgical option for the treatment of symptomatic diaphyseal osteonecrosis.

Take home message: Osteonecrosis must be considered in all patients receiving high dose or long term steroids for any indication. Intramedullary nailing can be a successful method of treating symptomatic diaphyseal osteonecrosis of long bones.

Transposition of a pancreas transplant from the bladder to the terminal ileum twenty years after combined allogenic kidney-pancreas-transplantation

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Introduction: During the first years of combined allogenic kidney-pancreas-transplantation bladder diversion of the exocrine pancreas secretion was used. After reporting urological and systemic complications it was switched to an enteric diversion with excellent results of pancreas function. Today enteric diversion of the pancreatic ductal secretion is the standard procedure. Nevertheless there are still patients alive with bladder diversion from the early years of transplantation with a good pancreatic function but loss of kidney function. This case describes such a patient and how we dealt with the problem.

Case description: A 53-year old male patient presented with a progressive renal failure twenty years after combined allogenic kidney-pancreas-transplantation with bladder diversion of the exocrine pancreas secretions. Urine excretion was declining with a pre-dialysis renal failure. Still the pancreas transplant was working properly without the need of insulin therapy. We therefore carried out a separation of the graft duodenocyostomy and re-established diversion by a side-to-side graft duodenal-recipient ileal anastomosis. This was done by a 2-layer hand sewn technique. Bladder catheter, drainage near the bladder and drainage near the anastomosis were removed after 5, 8 and 10 days respectively. The pancreas showed proper function without the need of insulin therapy. The patient was released from hospital 14 days after the operation.

Results and Conclusions: Transposition of a pancreas transplant from the bladder to the terminal ileum twenty years after primary transplantation is technically possible. In this case it was also reasonable in order to protect the bladder from the aggressive pancreatic ductal secretion. Because of the declining urine excretion due to progressive failure of the kidney transplant the exocrine secretion was not properly diluted anymore with the risk of hematuria, lower urinary tract infections, reflux-associated pancreatitis and transitional cell dysplasia. These conditions could limit the opportunity for the patient for a second kidney donation.

Take home message: Transposition of a pancreas transplant from the bladder to the terminal ileum twenty years after primary transplantation is technically possible and reasonable to offer the patient a chance for a second kidney donation.

Pancreatic malignancy presenting as duodenal ulcer (D1)

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Introduction: Malignant ulcer at the duodenal bulb is extremely rare. The commonest cause of ulcers here is peptic ulcer disease. Therefore the routine biopsy of ulcer at D1 is not routinely recommended, to avoid complications.
**Case description:** An 83-year-old lady presented to the surgical outpatient clinic with upper abdominal pain radiating to left side of her chest. She had recent history of significant weight loss, approximately 22 kg. Abdominal examination was unremarkable.

**Results and Conclusions:** Haemoglobin and Liver Function Tests were normal. Gastroscopy showed one large 3 cm 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 2.9x3.7 cm 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.

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**Changing standards in the treatment of desmoid tumors?**

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**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 4 cmx2 cmx3 cm 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.

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**A case report of metastatic renal oncocytoma**

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**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 24 hour 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 7.5x5.7 cm, extending to the renal pelvic with compression of the vascualar 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 chromosome 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**