ALK-Negative Anaplastic Large Cell Lymphoma Presenting As Recurrent Groin Lump

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Abstract

This case report describes the presentation of an uncommon Non-Hodgkin Lymphoma in a 72 year old male - ALK negative anaplastic large cell lymphoma, a recent entity formally recognized by the World Health Organisation (WHO) classification in 2016. Initially thought to be an inguinal hernia, the patient was diagnosed with a pathological inguinal lymph node at time of procedure with histopathology later confirming the diagnosis. Anaplastic large cell lymphoma is topical within surgical disciplines currently due to a recently identified association with breast implants.

CASE REPORT

A 72 year old male was referred to the Emergency Department with an incarcerated right inguinal hernia of 24 hours duration. The patient was known to have a groin lump and presented 6 months prior. The patient was referred to a surgeon and offered an open right inguinal hernia repair. Surgical intervention was declined as the groin lump presented intermittently and the patient was otherwise asymptomatic without any complications of hernia. The patient was systemically well without B symptoms of unintentional weight loss or night sweats. The patient was retired but worked on a hobby farm he owned with his wife. There was no preceding history of trauma. Past medical history included ischaemic heart disease, multiple sclerosis currently in remission without any significant neurological deficits, hypertension and dyslipidaemia. The patient was an ex-smoker with a 60 pack year history, had undergone a previous open appendicectomy, and reported a family history of prostate cancer but no other malignancy.

On presentation to the Emergency Department the patient was afebrile and hemodynamically stable without any obstructive symptoms. The abdomen was soft and non-tender. There was a painful right-sided groin lump present which was indurated with an overlying erythema. This was not reducible and did not have a cough impulse. Clinically there was no fluctuance or any inguinoscrotal extension and no obvious portal for infection was present. No other hernias were clinically evident. A CT abdomen/pelvis was performed which demonstrated a collection measuring 40x28x56 mm in the right inguinal region. Multiple liver lesions were visualised on CT of uncertain significance without any suspicious lymphadenopathy. An incision and drainage was performed however intraoperatively only a single 6cm necrotic superficial inguinal lymph node was found. The lymph node was excised and sent for histopathology and flow cytometry. The patient was well post operatively and discharged with outpatient follow up.

The patient was seen in surgical outpatients 2 weeks post discharge. The patient was systemically well with no concerns. The wound was healing well without any infective signs or significant underlying seroma/haematoma. There was no clinically evident cervical chain, supraclavicular, axillary or inguinal lymphadenopathy. Histology had confirmed the lymph node diagnostic for ALK negative Anaplastic Large Cell Lymphoma (ALK - ALCL). The results were conveyed to the patient and he was referred to haematology for further opinion and management.

DISCUSSION

ALCL refers to a group of Non-Hodgkin's Lymphomas (NHL) of T cell lineage that are characterised by the expression of CD30. Morphologically ALCL may be further distinguished from other CD30 receptor positive

lymphoma's through the presence of 'hallmark cells'; large polymorphic lymphoid cells containing abundant cytoplasm and horseshoe or kidney-shaped nuclei with an intermediate nuclear: cytoplasmic ratio and eosinophilic perinuclear clearing 4 5 7. ALCL comprises 2-3% of all adult NHL, 10 – 20% of childhood lymphomas and 12% of T cell NHL 2 4.

ALCL is topical within the field of surgery currently due to a recently demonstrated association in patients with breast implants. Breast implant-associated ALCL is now recognised as a separate entity and the subject of focus in recently published National Comprehensive Cancer Network (NCCN) consensus guidelines 3 9.

ALCLs may be further classified as ALK positive and ALK negative ALCL due to the presence of a novel chimeric fusion protein, ALK. ALK is formed most commonly from a translocation involving the anaplastic lymphoma kinase (ALK) gene on chromosome 2p23 and the nucleophosmin (NPM) gene on chromosome 5q35 but other variant rearrangements may occur 1 5.

ALK negative ALCL usually presents in the 5th or 6th decade of life in adults where ALK positive ALCLs more commonly present in children and young adults. On diagnosis, patients with ALK negative ALCL usually present with advanced disease - stage III or IV and often report B symptoms. Extranodal sites are frequently involved and include the skin, soft tissue, bone, lung, liver and bone marrow. The central nervous system may be involved but this is less common 1 2 4 5 7. ALK negative ALCL is not as prevalent as ALK positive ALCL generally has a poorer prognosis with 5 year overall survival reported as 30 -49% compared to 70 – 86% in ALK positive ALCL1 4.

This case describes an unusual presentation of a recurrent groin lump initially thought to be an inguinal hernia but later confirmed to be a pathological lymph node. A hernia is a protrusion of viscus or other structure beyond the normal coverings of the cavity in which it is contained 6. Hernias are a common problem in general surgery. The true incidence of hernias is unknown but it is estimated that 5% of the population will develop an abdominal wall hernia of which the majority occur in the inguinal region 8. The natural history of a hernia is one usually consisting of slow progressive enlargement exacerbated by increased intraabdominal pressure. Careful physical examination is required to determine anatomical location and reducibility. Where diagnosis is unclear on history and physical examination imaging including ultrasound should be used to exclude other potential causes. A diagnosis of ALK –ALCL was made based on the histopathology.

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