Castleman's Disease- Incidental or Instrumental?

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Background

Castleman's Disease refers to a group of lymphoproliferative disorders which share a series of histopathological features but have varying aetiologies, clinical features and outcomes[1].

It may be classified as unicentric or multicentric depending or whether a single or multiple lymph node regions are involved. Castleman's disease in association with HHV-8 is well described. The pathogenesis of HHV-8 negative Castleman's disease is poorly understood and there are no known infective precipitants.

References:

1. Dispenzieri A, Fajgenbaum DC. Overview of Castleman disease. Blood. 2020;135(16):1353-1364. doi:10.1182/blood.2019000931

2. Cronin DMP, Warnke RA. Castleman Disease: An Update on Classification and the Spectrum of Associated Lesions. Advances in Anatomic Pathology. 2009;16(4):236-246. doi:10.1097/PAP.0b

*3.https://www.pharmacodia.com/yaodu/html/v1/biologics/914101ec47c52b48a7b6ccc6f5a76f1f.*html

Clinical Case

A 54 year old man was referred to outpatient clinic 12 months post revision of a left total hip replacement. He reported left hip pain and had bulky left inguinal adenopathy on examination.

C reactive protein (CRP) was mildly elevated at 23.8. An aspirate of the hip was culture negative. 16s PCR (polymerase chain reaction) as well as specific staphylococcal and streptococcal PCR were negative but MEC A gene was detected in the sample. He underwent a further aspirate which again was culture negative and all PCR testing was negative.

In the absence of definitive evidence of prosthetic joint infection he proceeded to undergo a lymph node biopsy. This demonstrated changes consistent with HHV-8 negative, plasma cell variant Castleman's disease.

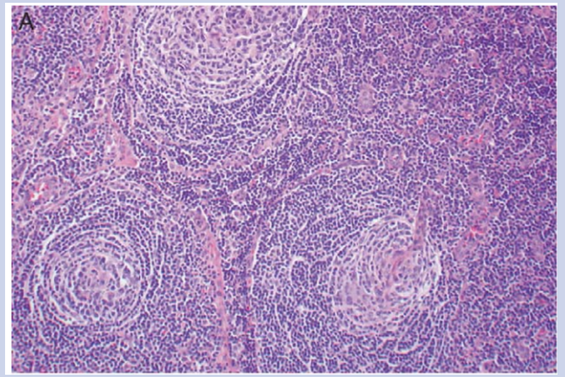

Figure A[2]

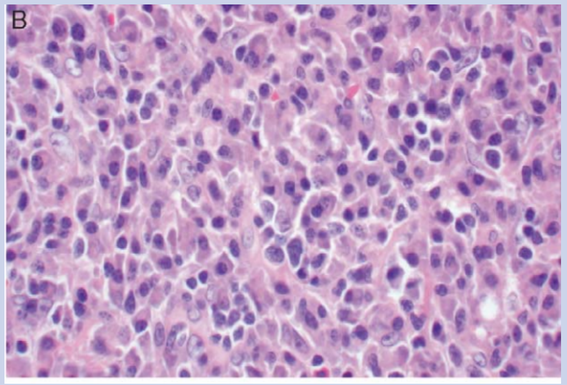

Figure B[2]

Representative of diagnostic findings in this case figure A demonstrates localized plasma cell Castleman's Disease with monotypic plasma cells and figure B demonstrates sheets of plasma cells- a key finding of this diagnosis[2]

Follow up/Discussion

The patient was subsequently reviewed by haematology and commenced on siltuximab- a chimeric monoclonal antibody which binds to interleukin-6

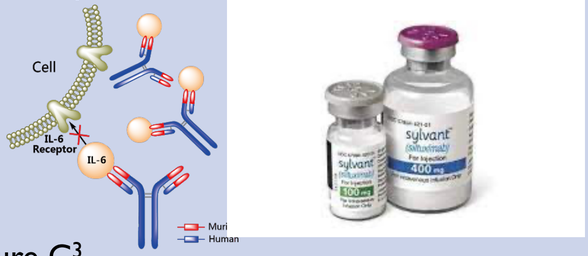

Figure C[3]

Despite treatment, the patient developed worsening left hip pain. Repeat CT scan demonstrated sclerotic change in the left femur with multiple collections in the soft tissues anterolateral to it. Repeat aspirate was again culture negative but MEC A gene was detected.

Distinguishing symptoms related to Castleman's from a low-grade prosthetic joint infection of the left hip is extremely challenging in this case. The use of IL-6 therapy further complicates this as CRP is no longer a reliable marker of systemic inflammation.

This case likely illustrates dual pathology- unicentric Castleman's presenting post operatively- a phenomenon not previously described in the literature and possible prosthetic joint infection of the left hip.