Soft Tissue Tumors: Lipoma: Chondroid lipoma

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Identity
Chondroid lipoma is a benign adipose tissue tumour that features clinical, morphologic, and genetic characteristics that are distinct from the common conventional lipoma.

Classification
Chondroid lipoma is a benign lipomatous neoplasm.

Disease
Chondroid lipoma

Epidemiology
Chondroid lipoma is an extremely rare benign adipose tissue tumour. Most tumours arise in adult women.

Figure 1. Lower and higher power fields (left and right respectively) of chondroid lipoma.
Chondroid lipoma primarily occurs in the subcutaneous or deep soft tissues of the proximal extremities and limb girdles, but this lesion may also arise in other sites including the distal extremities, trunk, and head and neck regions (particularly the oral cavity). This tumour typically presents as a painless, slow growing mass. Excision is curative; chondroid lipoma does not recur locally or metastasize.

Pathology
Chondroid lipomas range from 2 to 11 cm in size and are well delineated, often encapsulated lesions with yellow cut surfaces. Chondroid lipoma is composed of nests and cords of mature adipocytes and uni- or multivacuolated cells resembling lipoblasts embedded in a prominent myxohyaline matrix (Figure 1). Cells with eosinophilic, granular cytoplasms may also be seen. Hemorrhage and fibrosis are common with a prominent vasculature. Histologically, chondroid lipoma may resemble myxoid liposarcoma or extraskeletal myxoid chondrosarcoma. Immunostaining with S100 protein (mature adipocytes) is positive. Intracytoplasmic glycogen is highlighted with PAS stains. Alcian blue and toluidine blue stain the chondroitin sulfate substrate.

Cytogenetics
Chondroid lipoma is characterized by a t(11;16)(q13;p13) chromosomal translocation that results in fusion of the C11orf95 (11q13) and MKL2 (16p13.3) genes (Figure 2).

Genes involved and proteins
C11orf95
Location
11q13
DNA / RNA
The C11orf95 (chromosome 11 open reading frame 95) gene consists of 7 exons located within 6961 bp of genomic DNA.
Protein
C11orf95 codes for a 678 amino acid hypothetical protein of unknown function that exhibits expression in a wide variety of human tissues. This protein contains four C2H2 zinc fingers (classical zinc finger domain) with zinc ion binding molecular function.
Figure 3. Wild-type and fusion gene associated with the 11;16 translocation. A: Schematic of C11orf95, MKL2, and C11orf95-MKL2 fusion gene; solid bars represent coding exons, gray boxes are non-translated regions. B: Amino acid sequence at the breakpoint. C: Schematic and domain structure of the fusion C11orf95-MKL2 protein; the letters within the bars designate functional domains: zinc finger C2H2 domain (Znf), proline rich region (P), DNA-binding SAP domain (SAP), and coiled-coiled region (CC).

**MKL2**

**Location**
16p13

**DNA / RNA**
MKL2 is a member of the myocardin/megakaryoblastic leukemia gene family. The MKL2 gene contains 13 exons and spans more than 195 kb.

**Protein**
The MKL2 gene codes for a 1049 amino acid myocardin-like protein. MKL2, an SAP (SAF-A, acinus, and PIAS) DNA-binding domain containing protein, has been functionally implicated in chromatin remodeling in addition to serving as a transcriptional co-activator of SRF (serum response factor).

**Result of the chromosomal anomaly**

**Hybrid Gene**

**Description**
The 11;16 translocation results in fusion of exons 5 and 9 of the C11orf95 and MKL2 genes respectively.

**Fusion Protein**

**Description**
The C11orf95-MKL2 fusion gene codes for a transcript of 9127 nucleotides with an open reading frame of 3744 nucleotides. The chimeric transcript encodes a protein of 1247 amino acids that retains the SAP domain from MKL2.

**Oncogenesis**
The C11orf95-MKL2 chimeric transcript encompasses all putative functional motifs encoded by each gene. The C-terminal portion of the C11orf95-MKL2 chimeric protein contains a SAP DNA-binding domain, a coiled-coiled domain and a proline-rich region known to be present in transcription factors and oncoproteins. Additional studies must be conducted to determine the functionality of the C11orf95-MKL2 fusion oncogene in chondroid lipoma.

**References**


Ballaux F, Debiec-Rychter M, De Wever I, Sciot R. Chondroid lipoma is characterized by t(11;16)(q13;p12-13). Virchows Arch. 2004 Feb;444(2):208-10


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