

USP 6 Associated Tumors

Edited by: Dr.R.G.W.Pinto

Professor of Pathology

Former HOD

Former Dean Goa University

President Asian Society of Cytopathology

What is USP 6

Gene Rearranged

Tumors associated

Methods

Importance

USP 6

Ubiquitin Specific Protease 6

Gene Rearranged

Seen in

Aneurysmal Bone Cyst

Also seen in

Nodular Fascitis

Myositis Ossificans

Fibro osseous Pseudotumor of digits

Celluar Fibroma of tendon sheath

Clonal

Neoplastic

They have a Clinical and Histopathological overlap

USP 6 Gene Rearranged

Associated neoplasms

The Gene is on chromosome 17

Gene fusion

Fusion partner is usually CDH 11

USP 6 Gene fuses with CDH 11

Also COL1A1

MYH9

UBE2G

SNHG

USP 6 Gene. Rearranged in

ABC Aneurysmal Bone Cyst

Nodular Fascitis

Myositis Ossificans

Fibro osseous pseudo tumor of digits

Fibroma of tendon sheath

Tenosynovial Nodular Fascitis

And in

Soft Tissue ABC

Methods

CDH 11

USP 6 fuses with CDH 11

Fusion partners

USP 6.

Methods are

FISH

RT PCR

Sequencing

NGS

Dr Andre Oliveira showed that the USP 6 Gene Rearrangement
is seen in Primary ABC

Here USP 6 fuses with CDH 11

Rapid growth

Brisk mitosis

History of trauma

Andre Oliveira first

Later

Yahan Zhang et al

West China hospital

Sichuan Univ China

All ages

Young adults

Fibroblasts and Myofibroblasts proliferation

Osteoid

Osteoblasts

Multinucleate giant cells

Cartilage Metaplasia

Zonation

Myxoid areas

Extravasated RBCs

Inf cells

IHC positive

Smooth Muscle Actin

MSA

SATB2 in Bone Metaplasia

IHC Negative

Desmin -

S100 -

B Catenin -

MDM2 -

MIB1 is

1 % to 30 %

Treatment

Curettage

Resection

In Bone ABC Primary

The word Secondary ABC is no longer used

It is called ABC Like changes