Benign and reactive sarcoma like lesions

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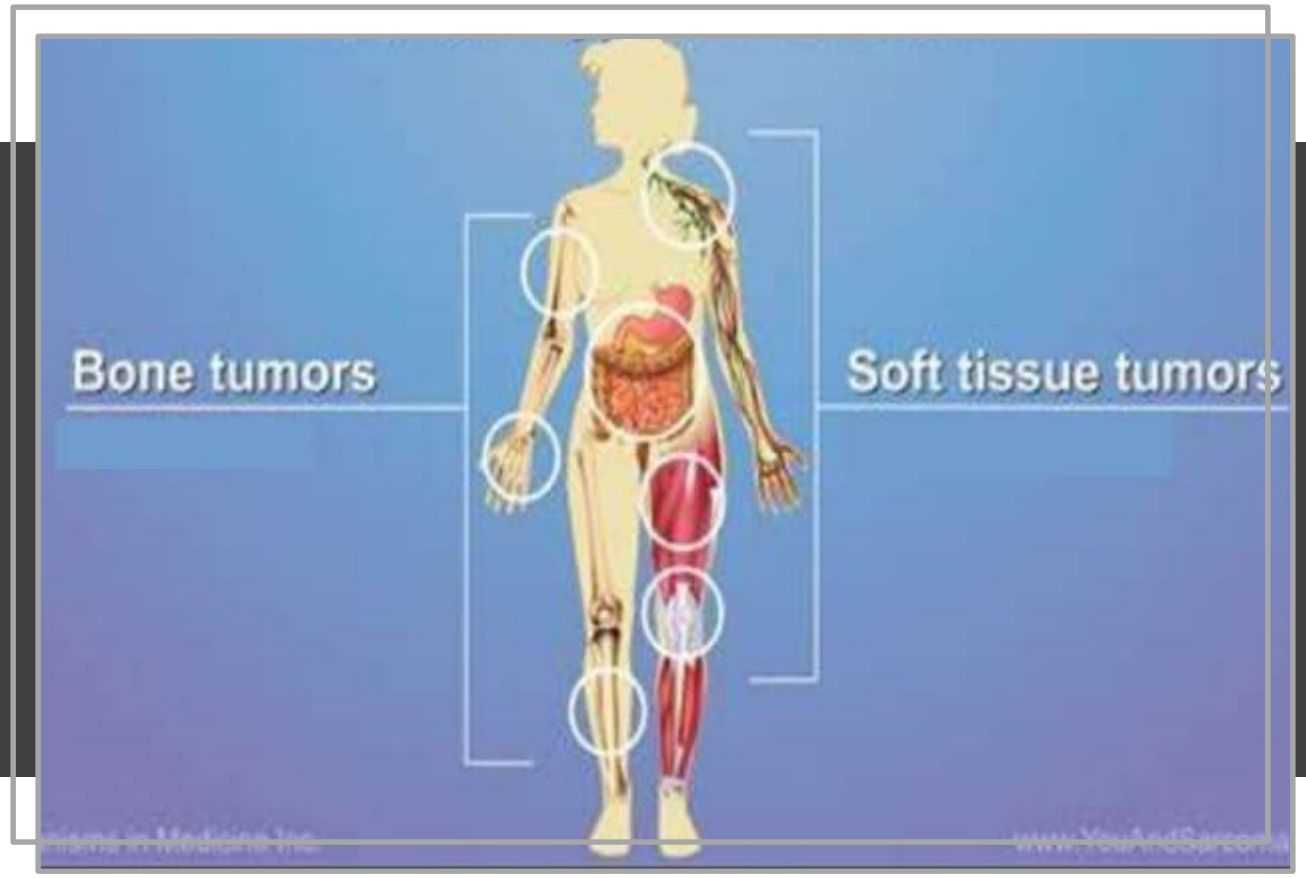
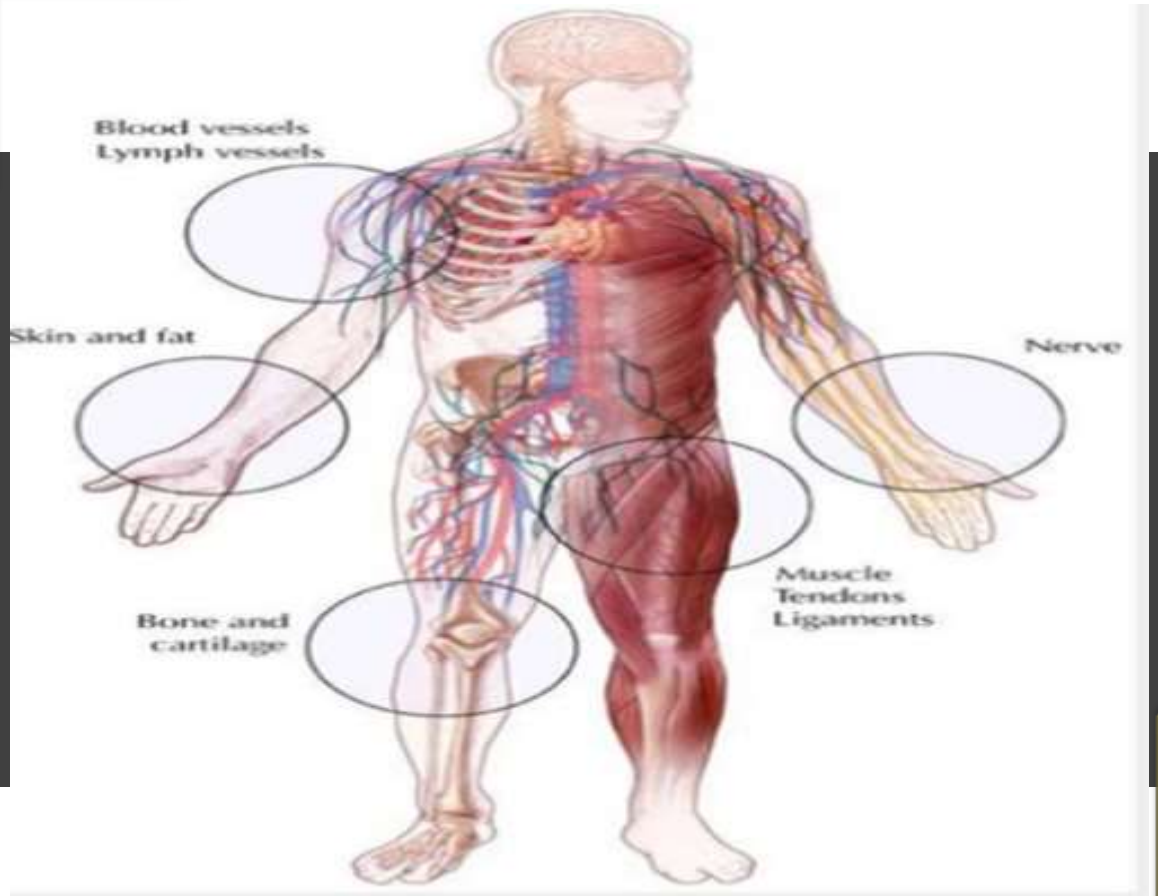
Term Sarcoma



Definition

Include 2 broad group of tumors which arise in bone or soft tissue.

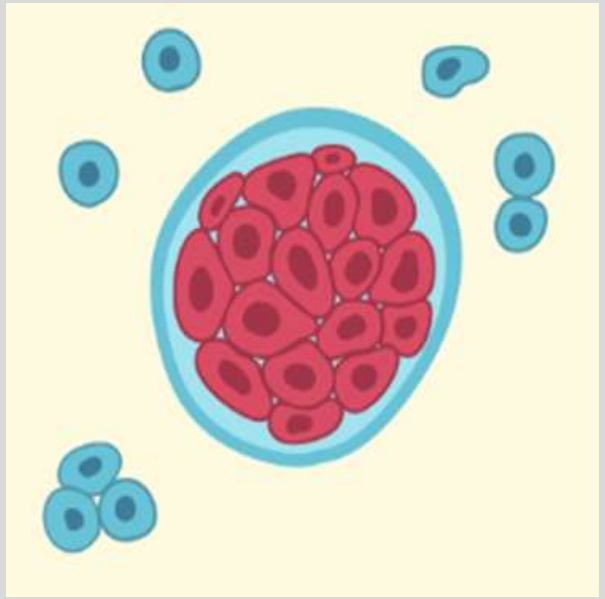
Soft tissue tumors



Site of origin

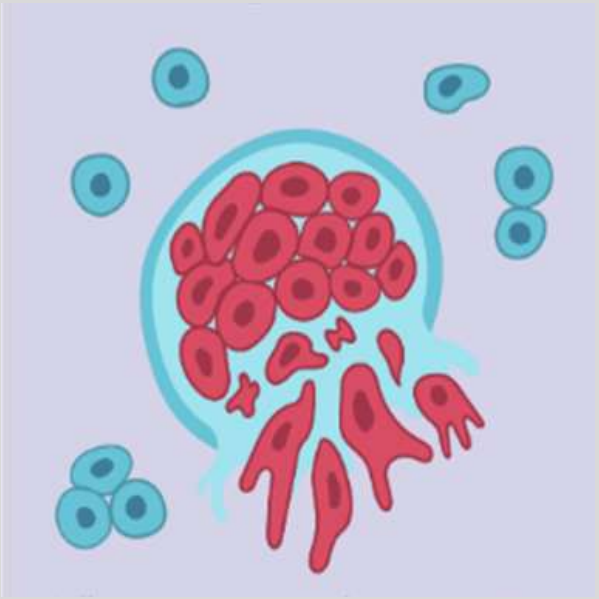
Occur in tissues that connect, support and surround other body structures. This includes muscle, fat, blood vessels, nerves, tendons and the lining of your joints.

Different soft tissue tumors.



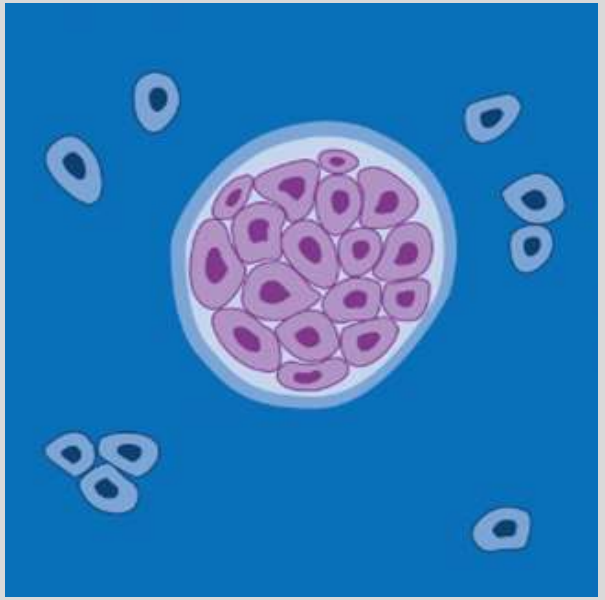
Benign

Cannot spread to other parts



Malignant

Cells spread to different parts of body are called soft tissue sarcoma



Intermediate

Grow and invade nearby organ but do not spread are **reactive and benign.**

Commonly seen Intermediate soft tissue tumors in clinical practice

(also called benign and reactive sarcoma like lesion)

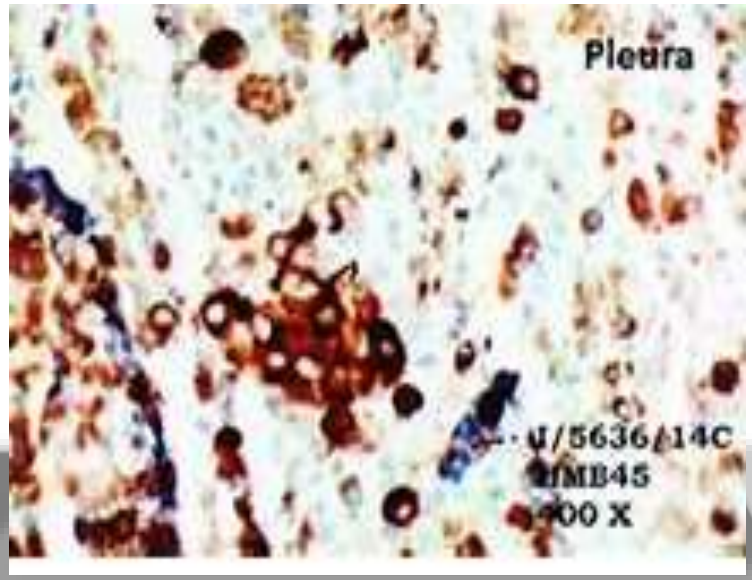
1. Dermatofibrosarcoma protuberans
2. Fibromatosis(Desmoid)
3. Solitary fibrous tumors
4. Atypical lipomatous tumor (ALT)
5. Infantile fibrosarcoma

Diagnosis of soft tissue tumor suspecting sarcoma like

History & PE

MRI is IOC

Biopsy & IHC



Important points while taking history of present illness of a swelling.

Duration

- It is important to note duration of swelling

Mode of onset & progress

- Started spontaneous or after trauma.
- Rapid or slow growing

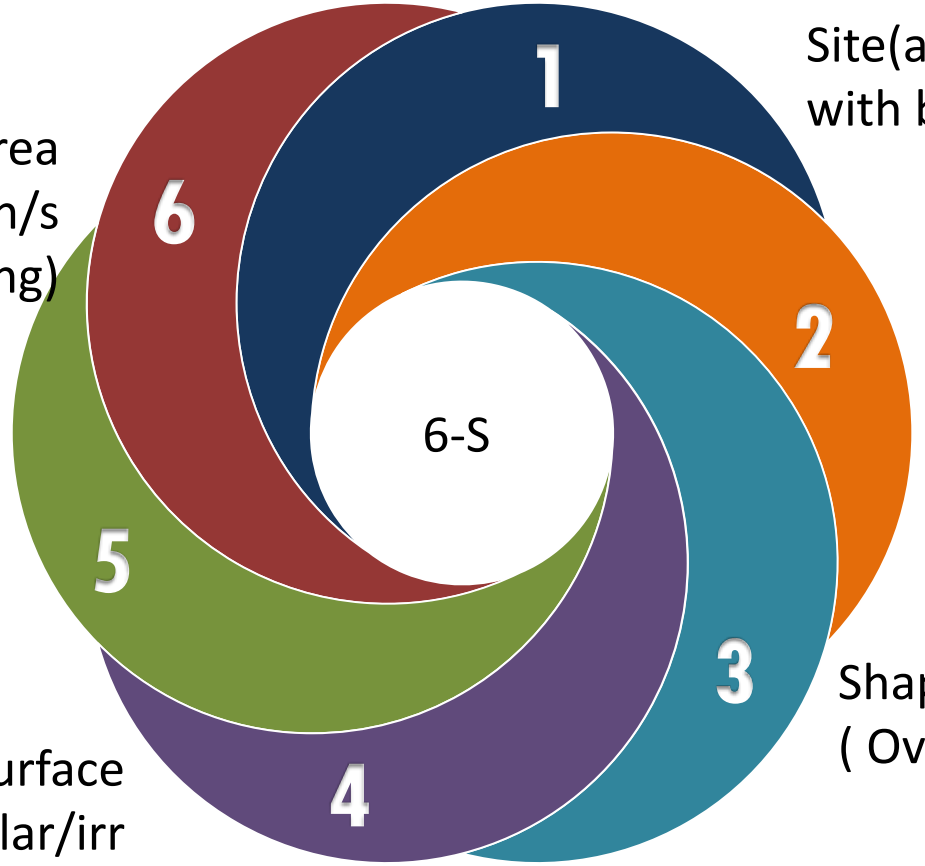
Pain & fever

- Any associated pain
- Relation of fever with swelling.

Other features

- Secondary changes
- Movement
- Loss of wt & appetite.

6- S of inspection of soft tissue swelling are



Site(also check its relation with bony land mark)

Size(vertical & horizontal dia.)

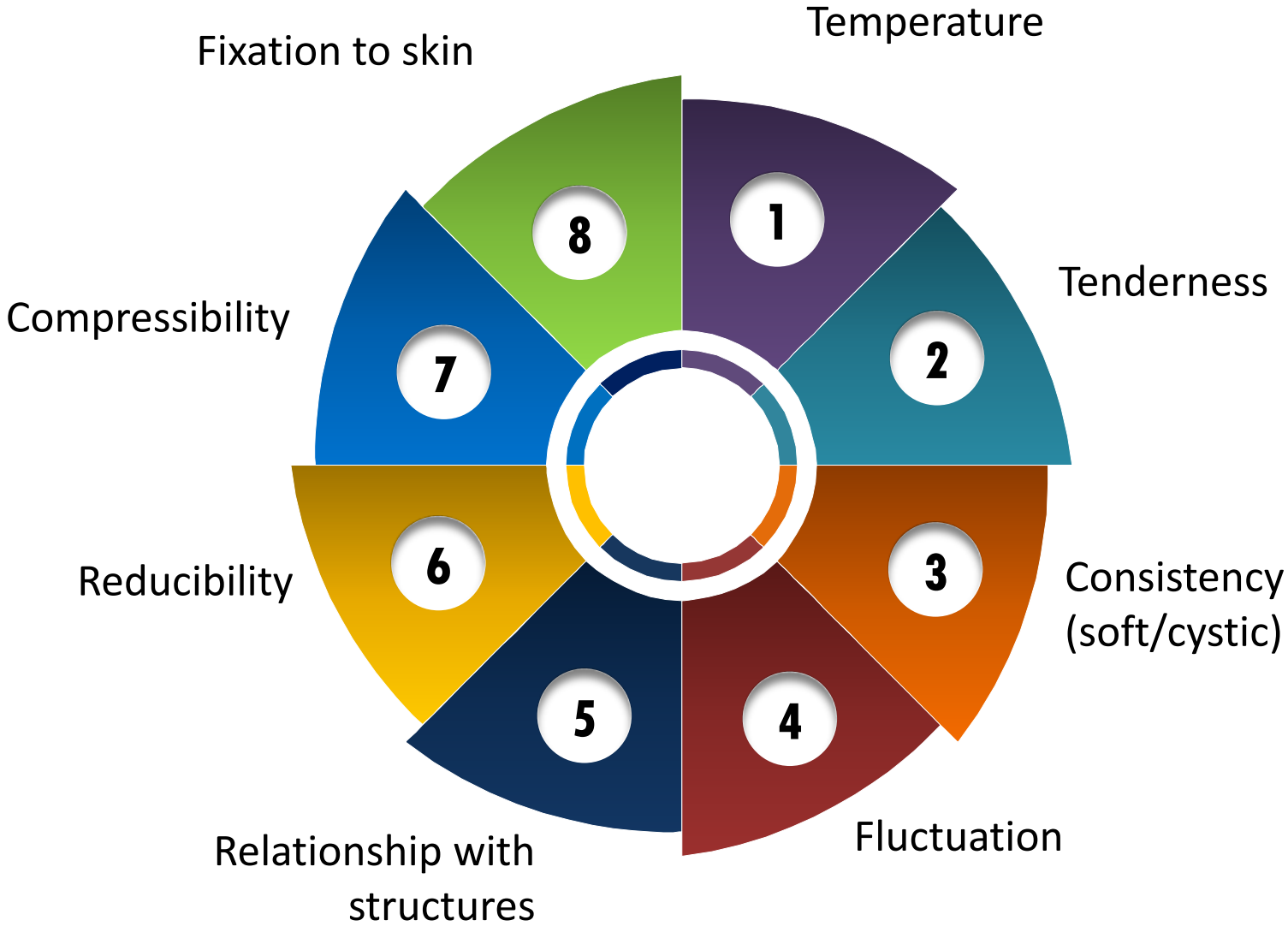
Shape
(Oval/globular/irregular)

Surface
(smooth/nodular/lobular/irregular)

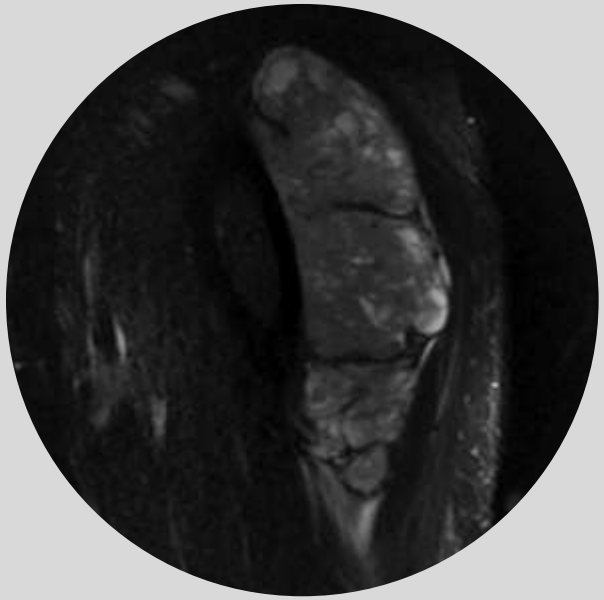
Skin
(tense/reddish/ulcer/discharge/fungating)

Surrounding area
(pigmentation/ulceration/swelling)

Defined order of palpation

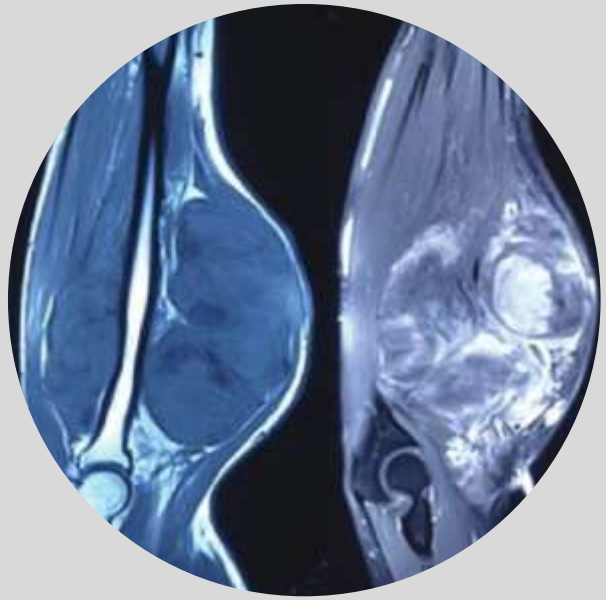


Features which are not seen on MRI in these tumors



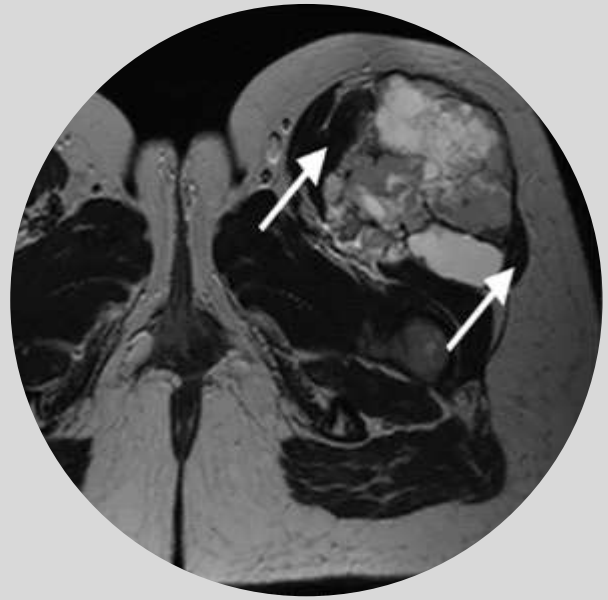
1

Complex heterogenous mass



2

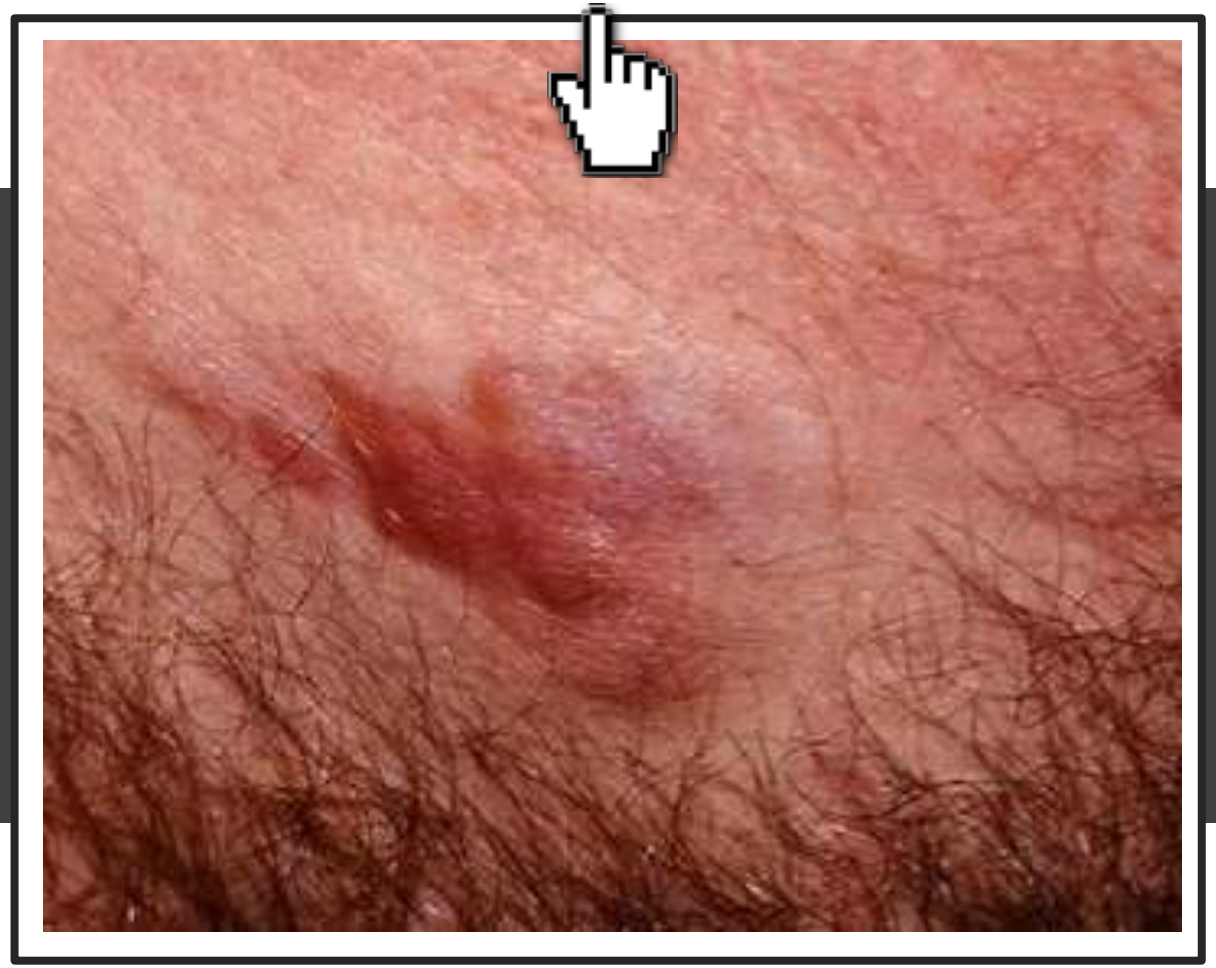
Post contrast enhancement



3

Presence of necrosis

Dermatofibrosarcoma protuberans



It slow-growing tumor of the fibrous tissue beneath the skin, usually in the trunk or limbs.

DFS

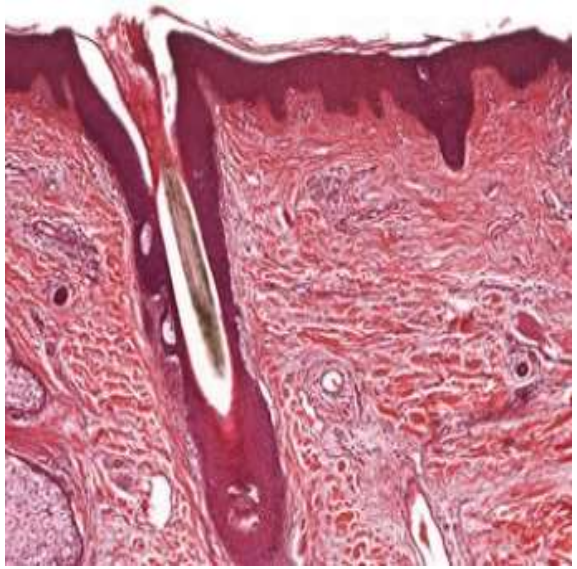
1-6%	Of all soft tissue tumors
30-50yr	Is most common age of presentation
85%	Occur in trunk and extemities

14-52% High rate of local recurrence after treatment

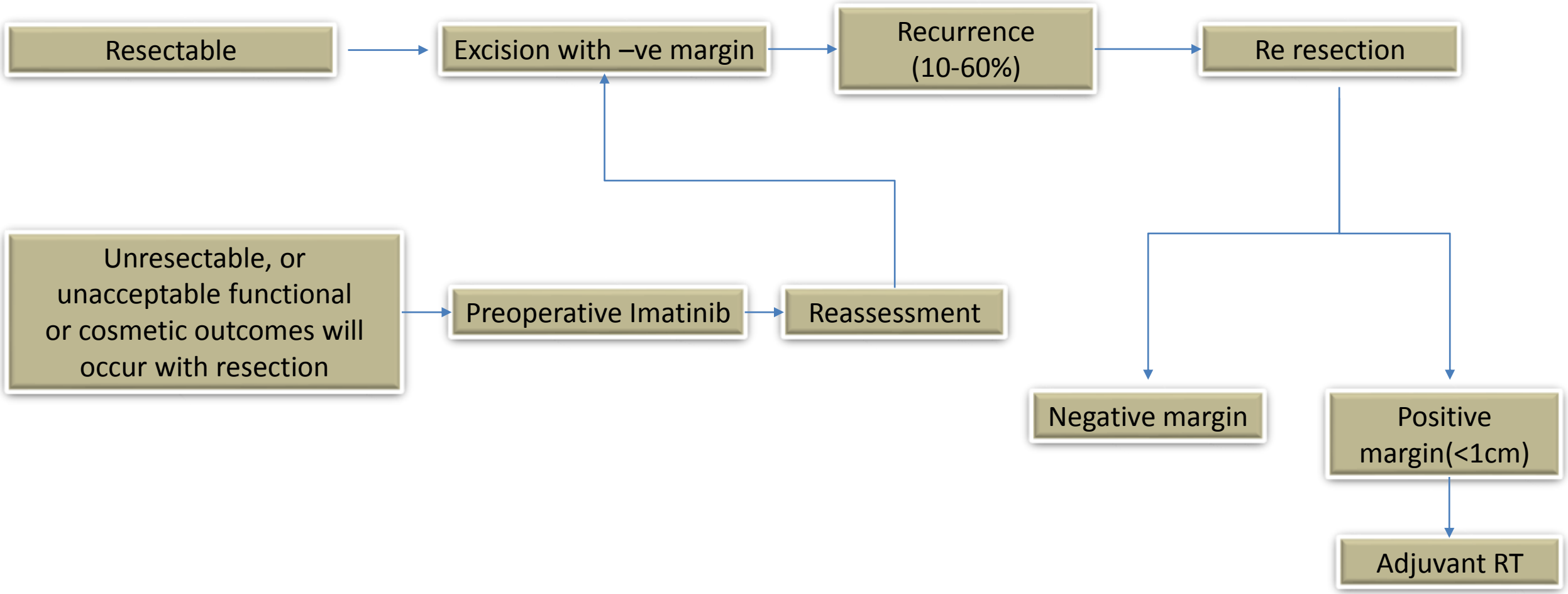
1-4% Risk of distant metastasis

99.1% 10 yr survival rate

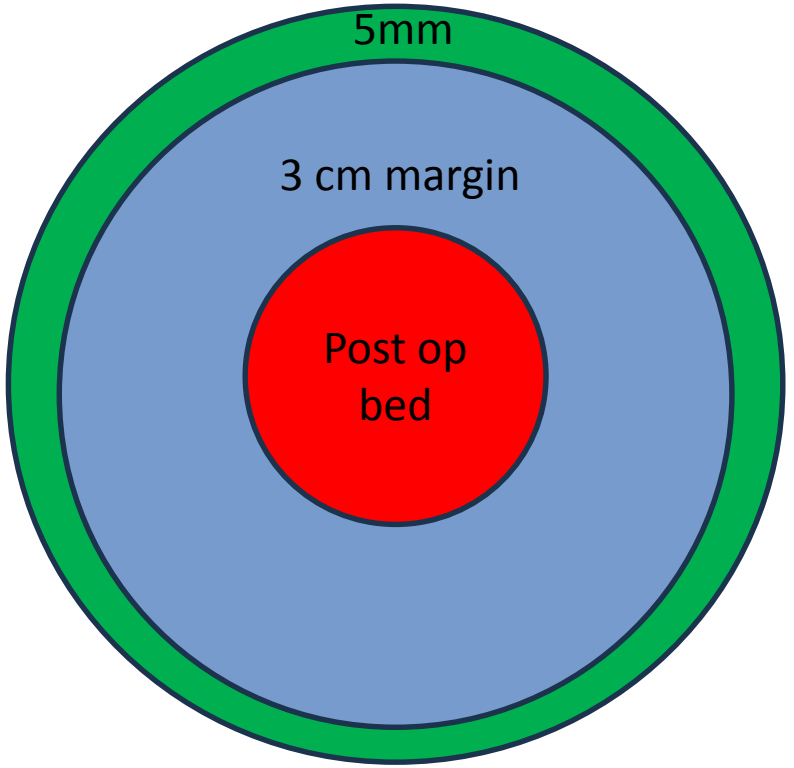
Pathological features of DFSP

Biopsy	IHC	FISH for equivocal case
		
Deeper subcutaneous layer	CD34 +ve S100 &factor XIIIa negative.	t(17;22)(q22;q13), is a hallmark

Management of biopsy confirmed DFSP



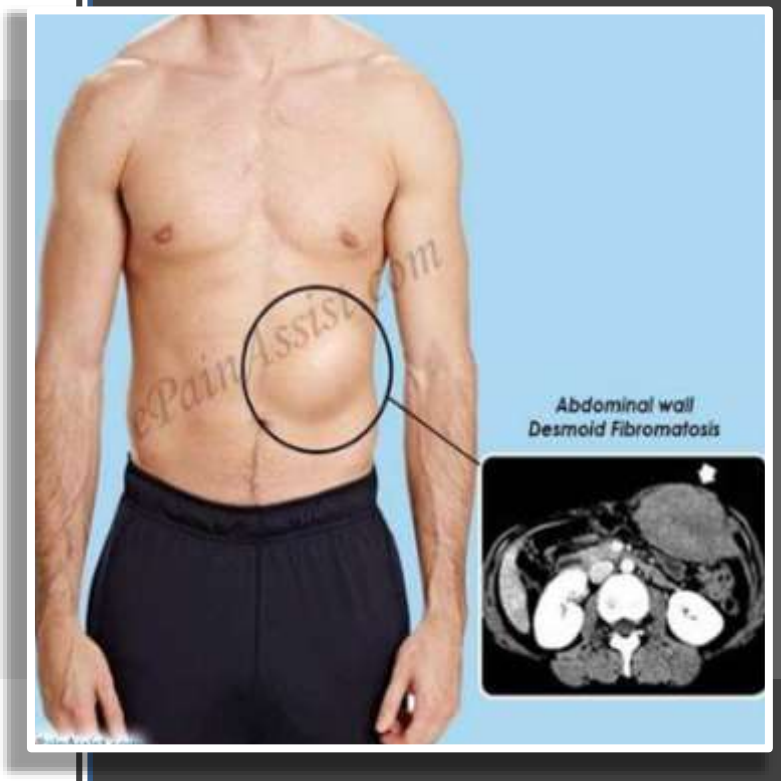
Adjuvant RT in margin positive DFSP



Dose is 66Gy to PTV

Desmoid Tumors (Aggressive Fibromatosis)

Clonal fibroblastic proliferation that arises in the deep soft tissues which grows slowly and steadily and rarely metastasis



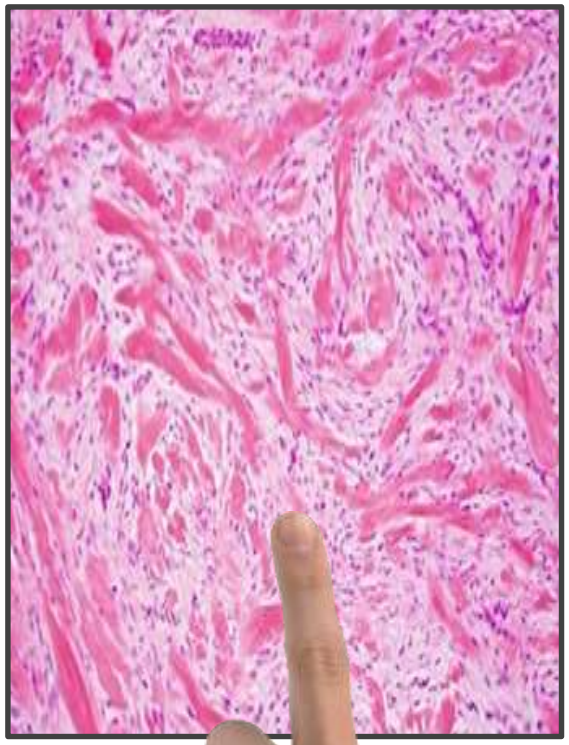
Desmoid tumors

3%	Of all soft tissue tumors
30-40yr	Peak age of presentation
7.5 cm	Median tumor size on presentation
10%	Occur with FAP and Gardner syndrome

Pathological features of desmoid

Microscopy

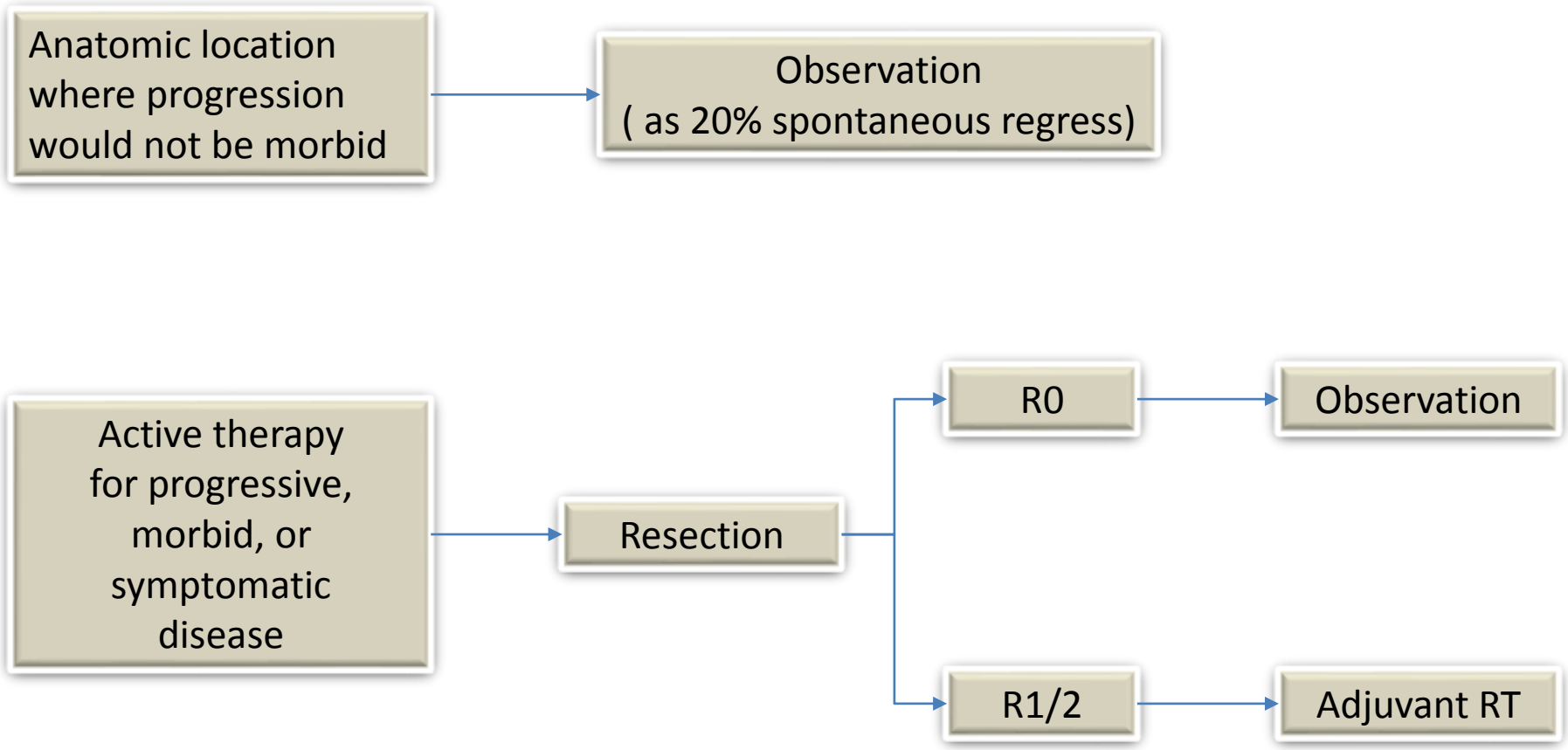
Proliferation of uniform spindle cells resembling myofibroblasts, in the background of abundant collagenous stroma and vascular network.
Hyperchromasia & nucli atypcia are absent



IHC

+ve for nuclear beta-catenin, vimentin,COX 2,androgen&estrogen receprtos

Management of desmoid



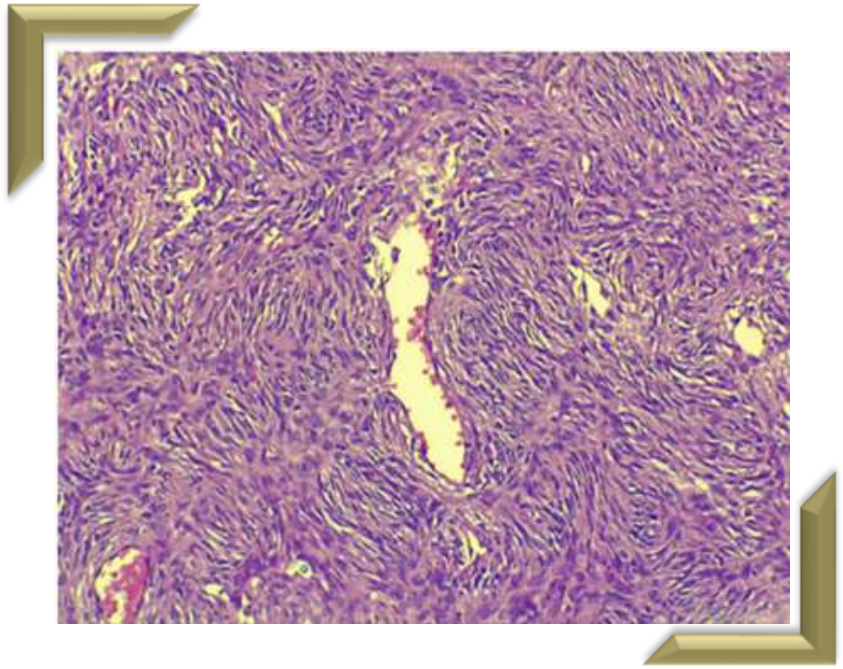
Solitary fibrous tumors (earlier called hemangiopericytomas)

Fibroblastic tumor which starts in the thigh, underarm, pelvis and also in pleura.

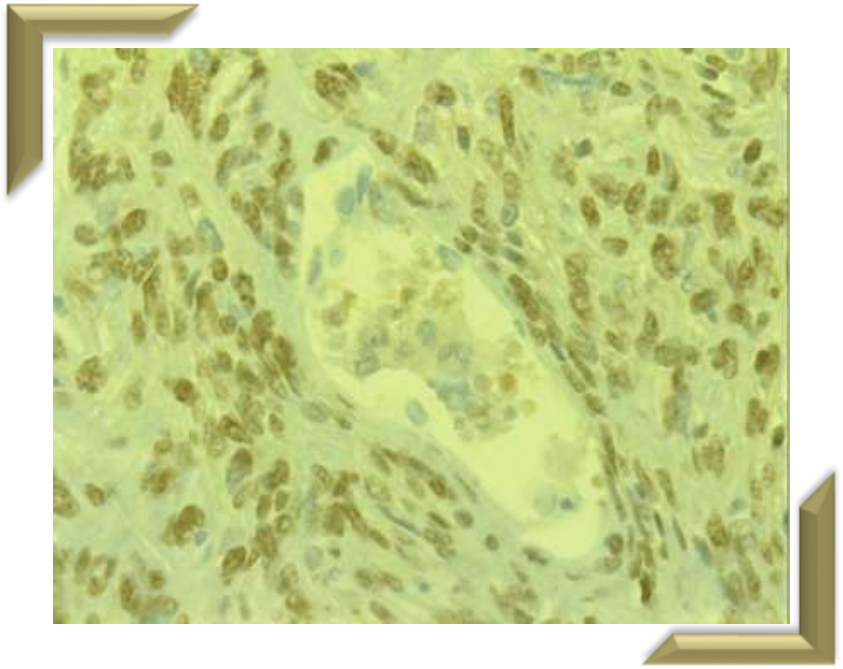
Solitary fibrous tumor

55yr	Median age of presentation
1/M	Age-adjusted yearly incidence
10%	Present with paraneoplastic syndrome (HOA)
Shows contrast enhancement being vascular	65%
Risk of metastasis	35-45%
Risk of relapse at 20 yrs	18%

Two histopathological features of solitary fibrous tumor

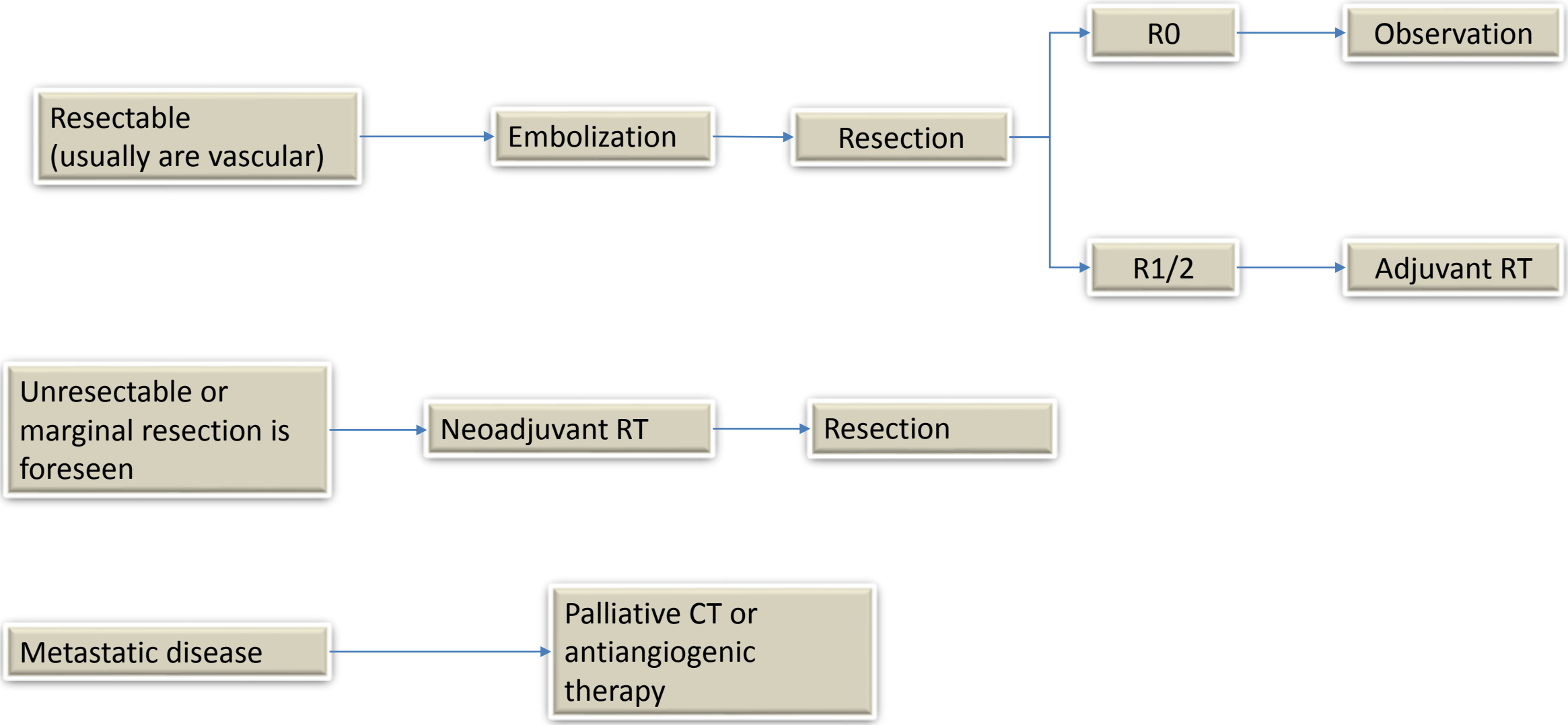


- Staghorn appearance



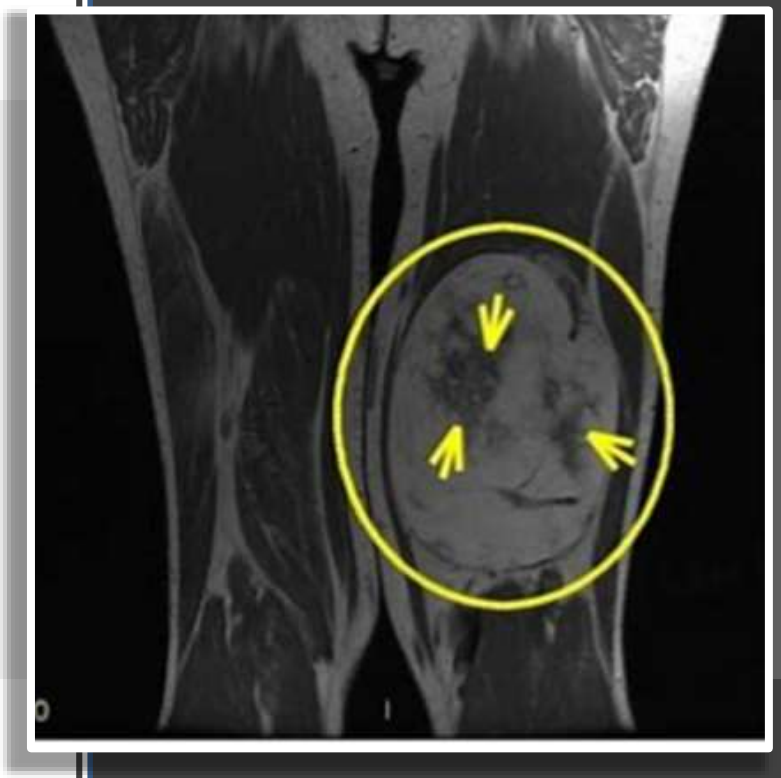
- STAT6 positive nuclear immunostaining

Management of SFT



Atypical lipomatous tumor (ALT)/Well-differentiated liposarcoma (WDLS)

Locally aggressive mesenchymal tumor from fat tissue



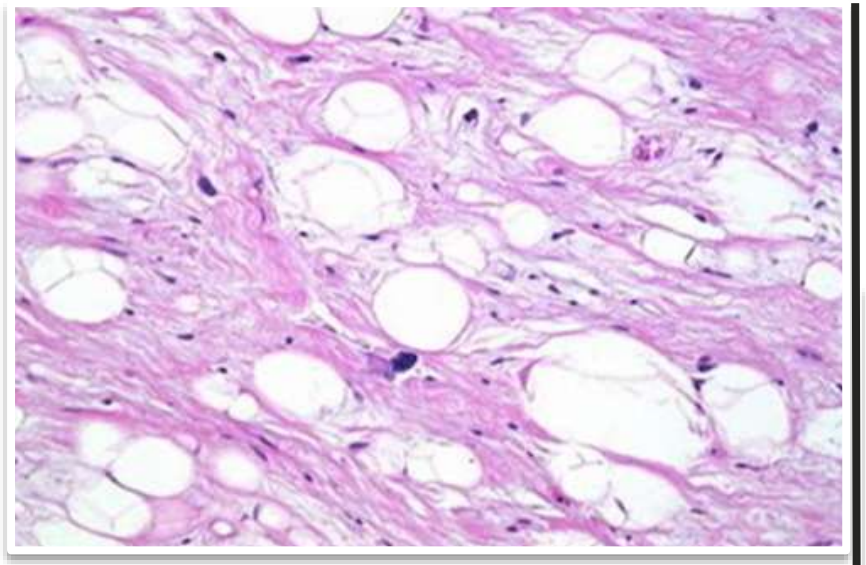
3 things of ALT are

Most common adipocytic malignancy
(40 - 45% of all liposarcomas)

40 - 60 years is peak age

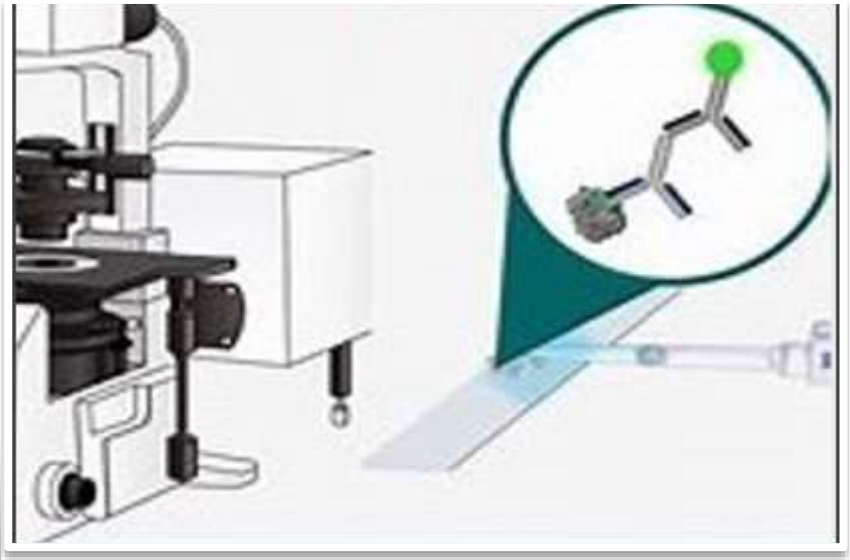
Do not metastasize unless they dedifferentiate

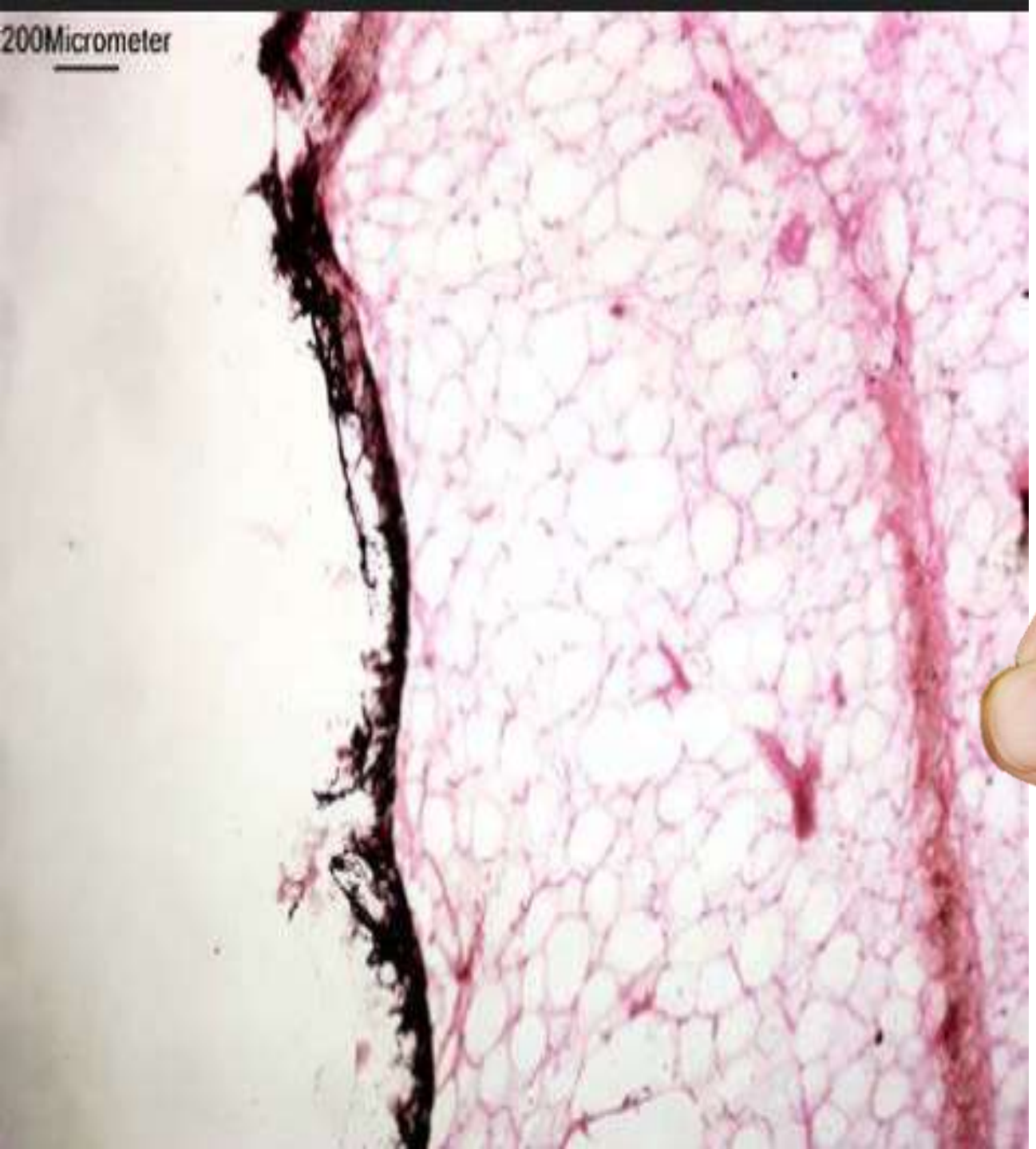
Pathologically



- Mature adipocytes and stromal cells with at least focal cytologic atypia

- MDM2, CDK4 and p16 together have 71% sensitivity and 98% specificity on IHC





Treatment

- TOC is complete excision with negative margin
- No role of adjuvant treatment

Infantile fibrosarcoma

1. 1% of all childhood cancers
2. Originates in the connective fibrous tissue found at the ends of long bones
3. Assoicated with ETV6 and NTRK3 gene fusion
4. Locally aggressive but rarely metastasizes
5. TOC is resection

Summary

1. Intermediate ST tumor are usually locally aggressive
2. MRI is IOC
3. Biopsy with IHC is cornerstone for diagnosis
4. Complete surgical removal with negative margin is TOC
5. Adjuvant RT is added if margins after resection are <1 cm
6. Chemotherapy has a palliative role in metastatic cases

Thank You

Contact Us For More Info

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