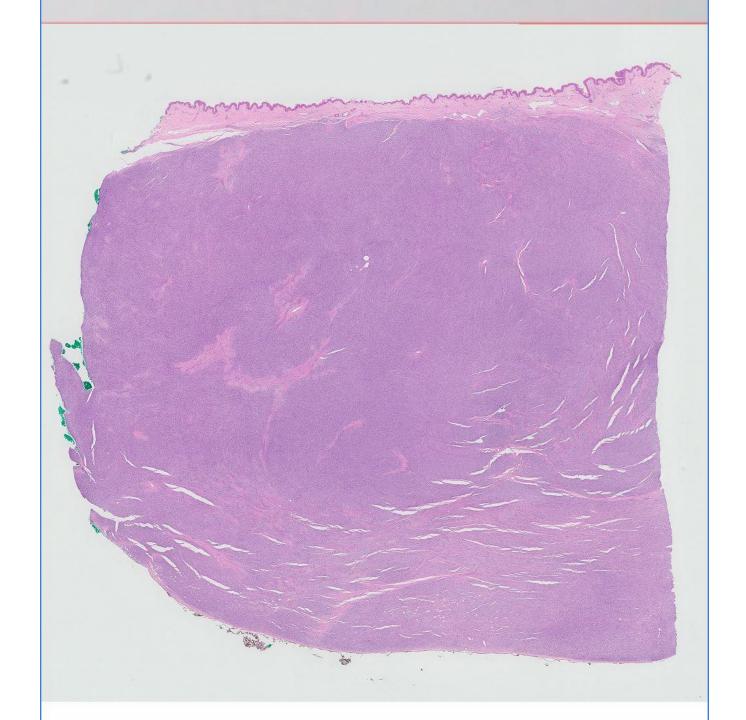
Case 7

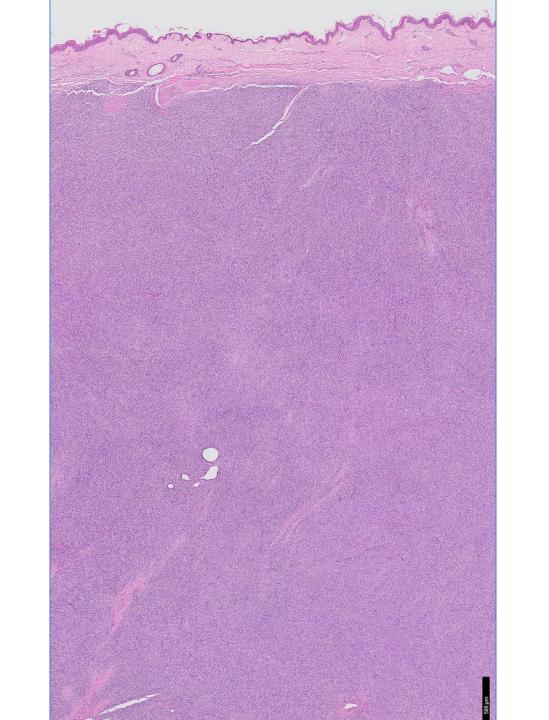
17 year old girl with a right breast lump, measuring close to 3cm.

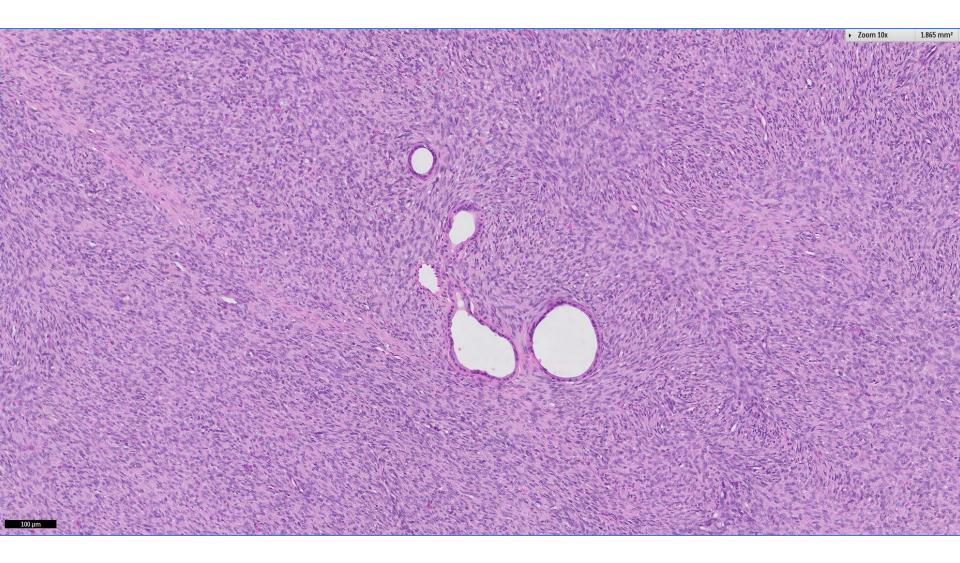
Excision biopsy.

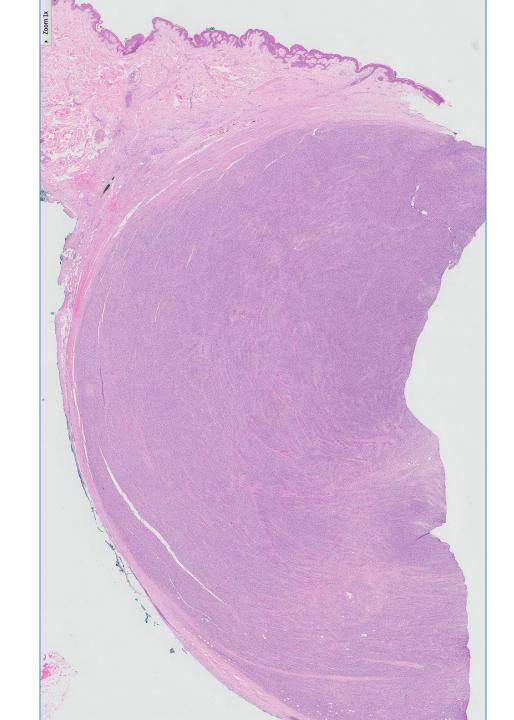


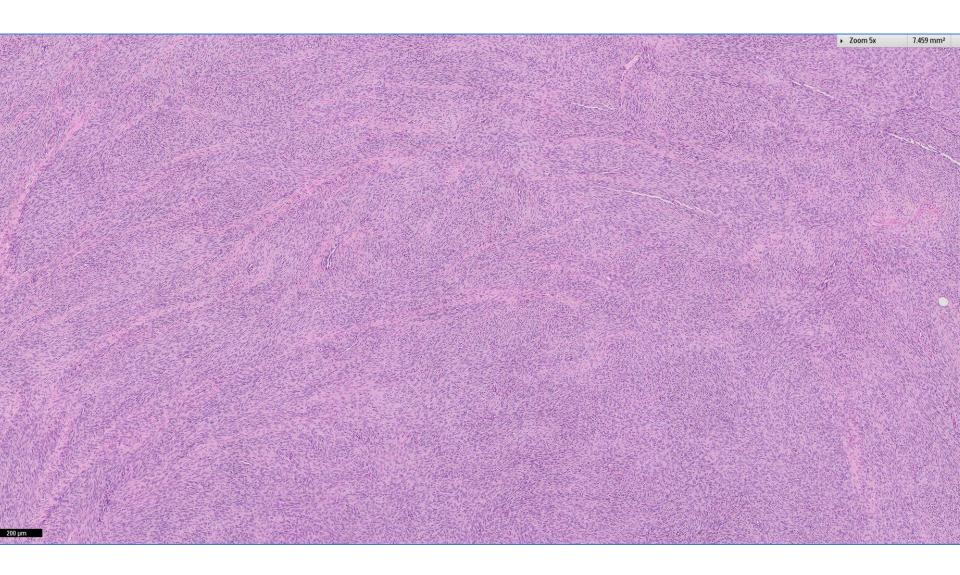




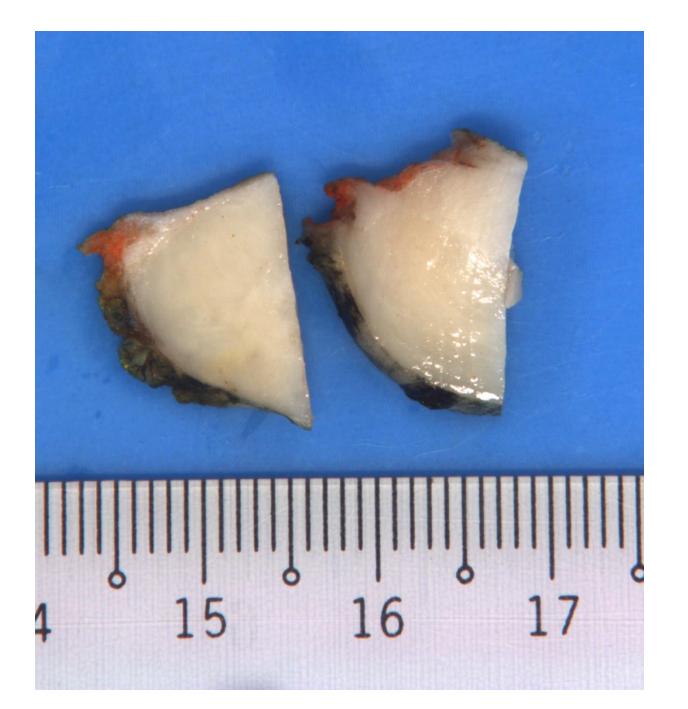


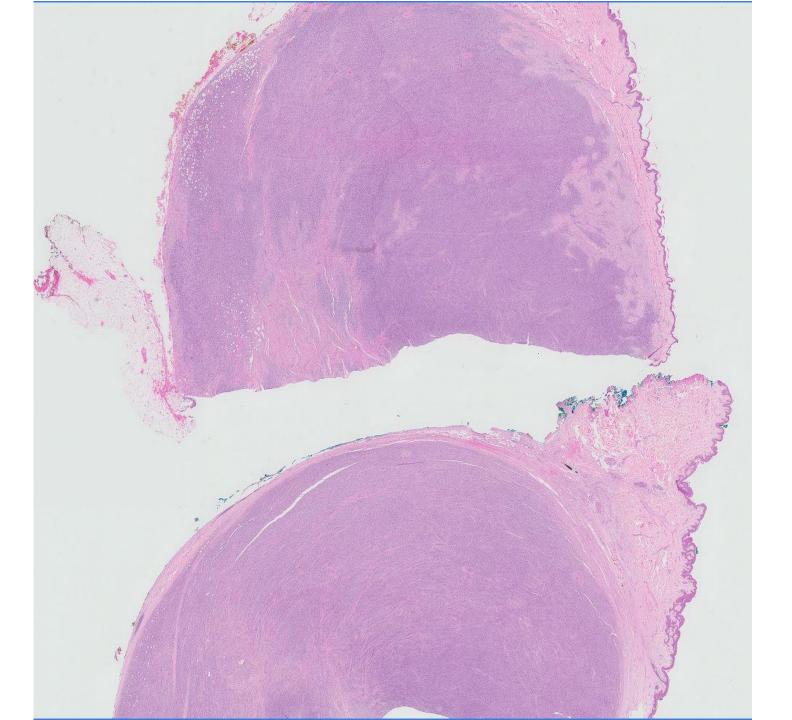


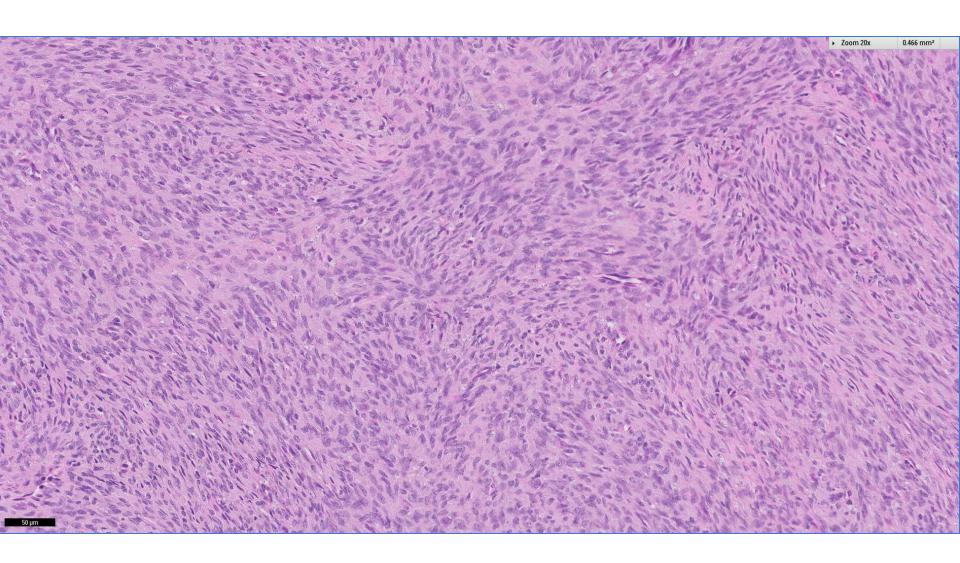


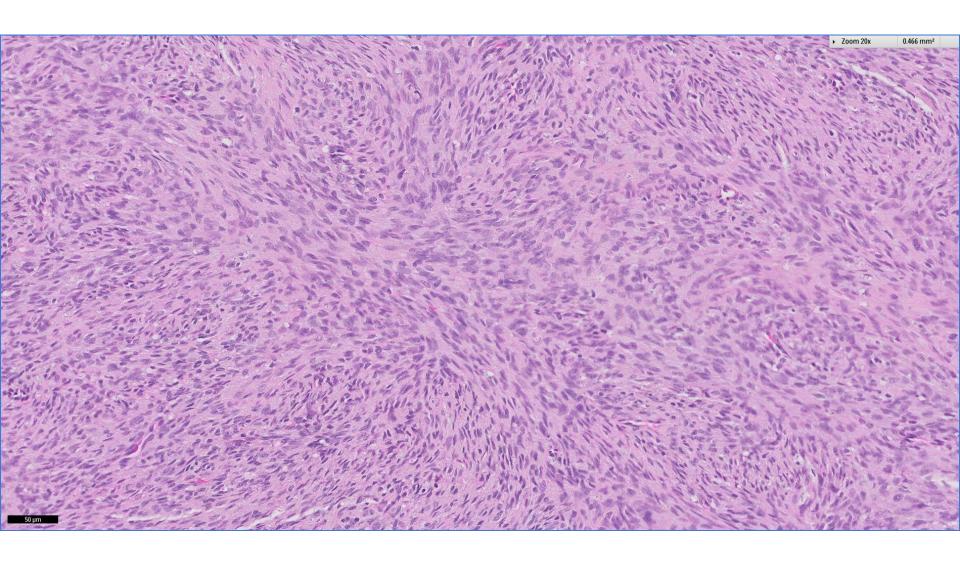


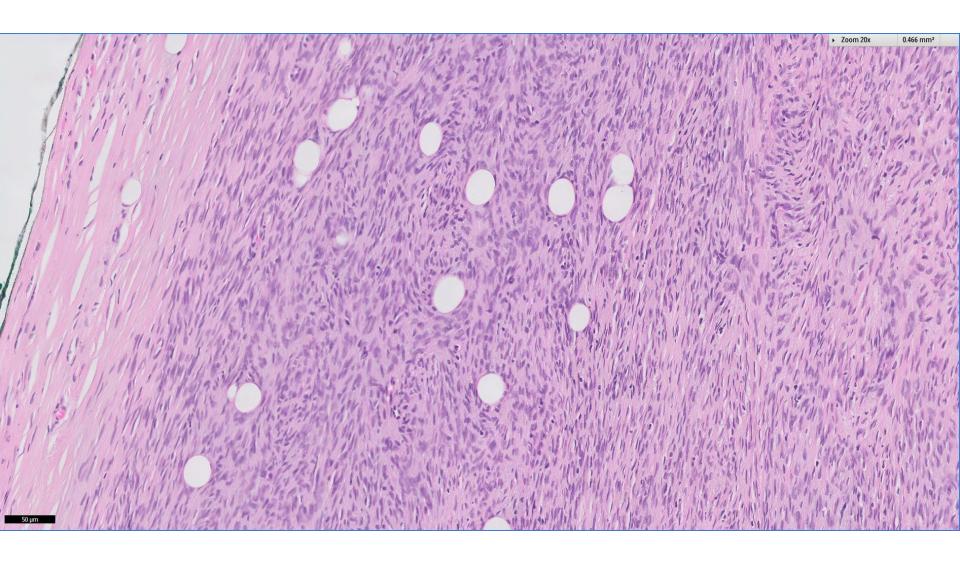


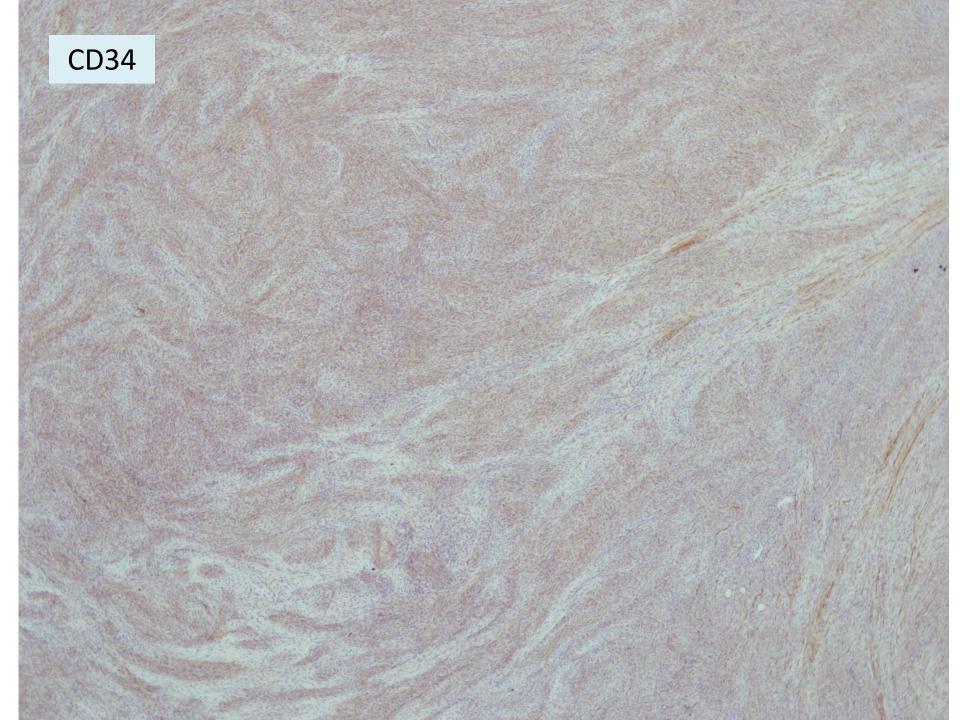


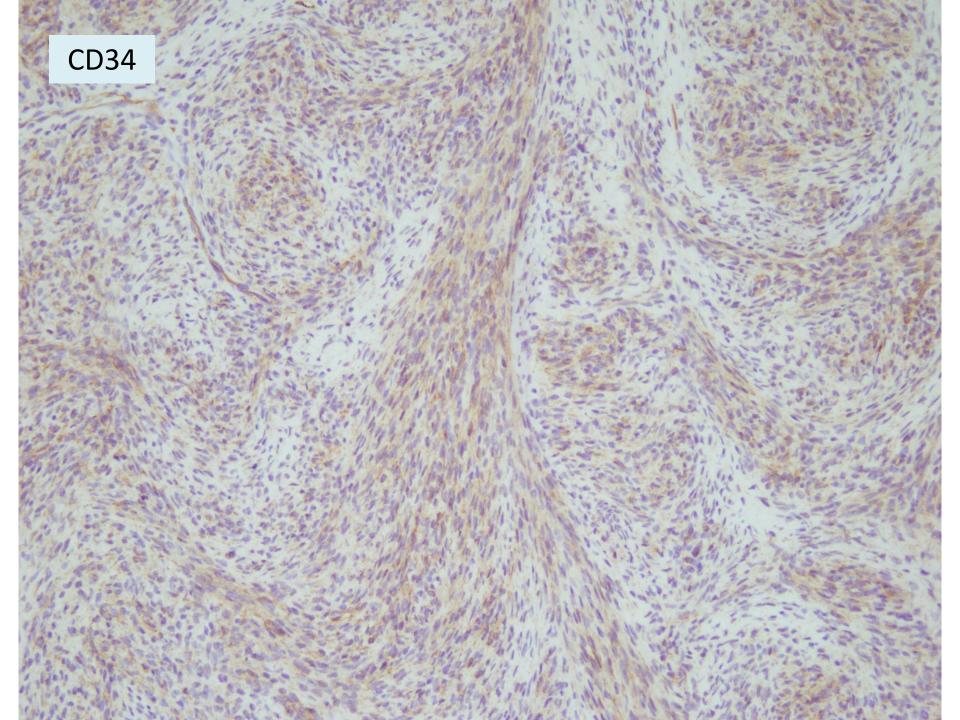


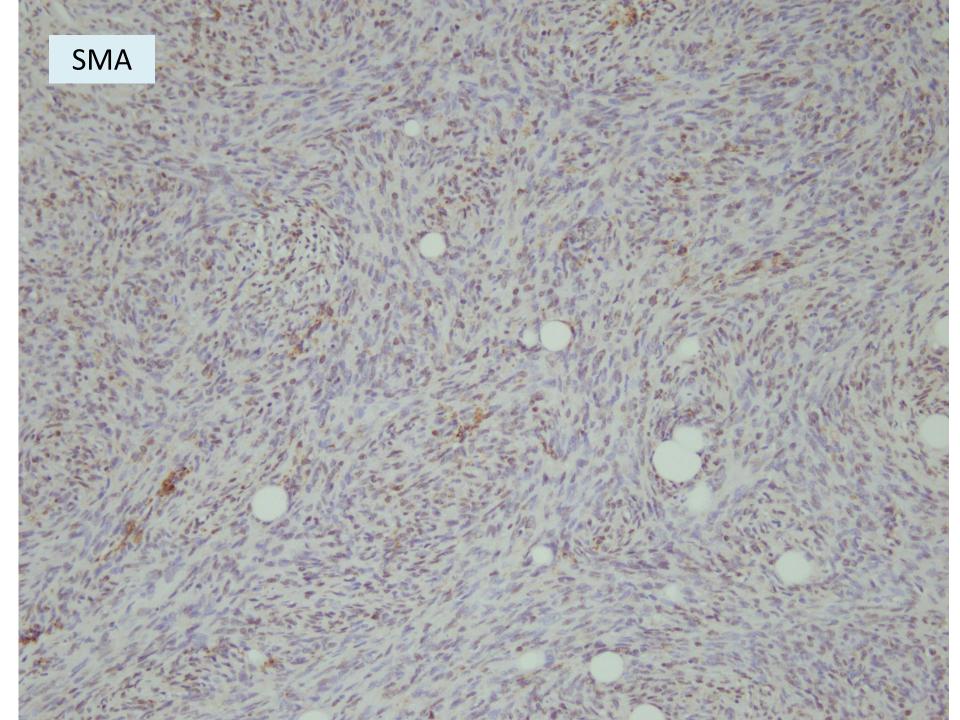












Other immunohistochemical results

- Bcl-2 diffusely & strongly positive.
- EMA, AE1/3 negative.
- Desmin negative.
- S100 negative.
- ER, PR negative.





Right breast lump, excision:

Dermatofibrosarcoma protuberans





Dermatofibrosarcoma protuberans

- Tumour arising from the skin and subcutaneous tissue.
- Not a primary breast neoplasm, but may present clinicoradiologically as a breast mass.
- Occurs at any age group, average age is 40 years.
- Immunohistochemistry:
 - CD34, vimentin positive.
 - S100, actin, desmin, CD31, cytokeratin, FXIIIA, EMA,
 HMB45, ER, PR, CD31, CD99, c-kit negative.





Dermatofibrosarcoma protuberans

- Genetic studies show characteristic translocation t(17;22)(q22;q23) in which the COL1A1 and PDGFB genes form a fusion gene.
- Fusion gene can be detected by chromosome analysis, FISH, RT-PCR (products of the fusion gene).





Dermatofibrosarcoma protuberans: treatment & prognosis

- Recurrent potential if not completely resected.
- Difficult to achieve negative margins due to the deep penetration of lesional cells into the surrounding tissue.
- Mastectomy is necessary for large bulky tumours; local excision for smaller lesions.
- May evolve into fibrosarcoma:
 - Development of herringbone growth pattern.
 - Increased cellularity.
 - Increased mitotic activity.
 - Necrosis.
 - Loss of CD34 reactivity.





Dermatofibrosarcoma protuberans: treatment & prognosis

- Recurrences usually occur within a few years, but can happen within 6 months or after decades.
- No role for adjuvant radiation treatment.
- Imatinib mesylate, an inhibitor of the PDGFR associated tyrosine kinase, may be useful in treating advanced cases.





