

Desmoid Tumors in Pediatrics

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Hope lives here.



Desmoid Tumors in Pediatrics

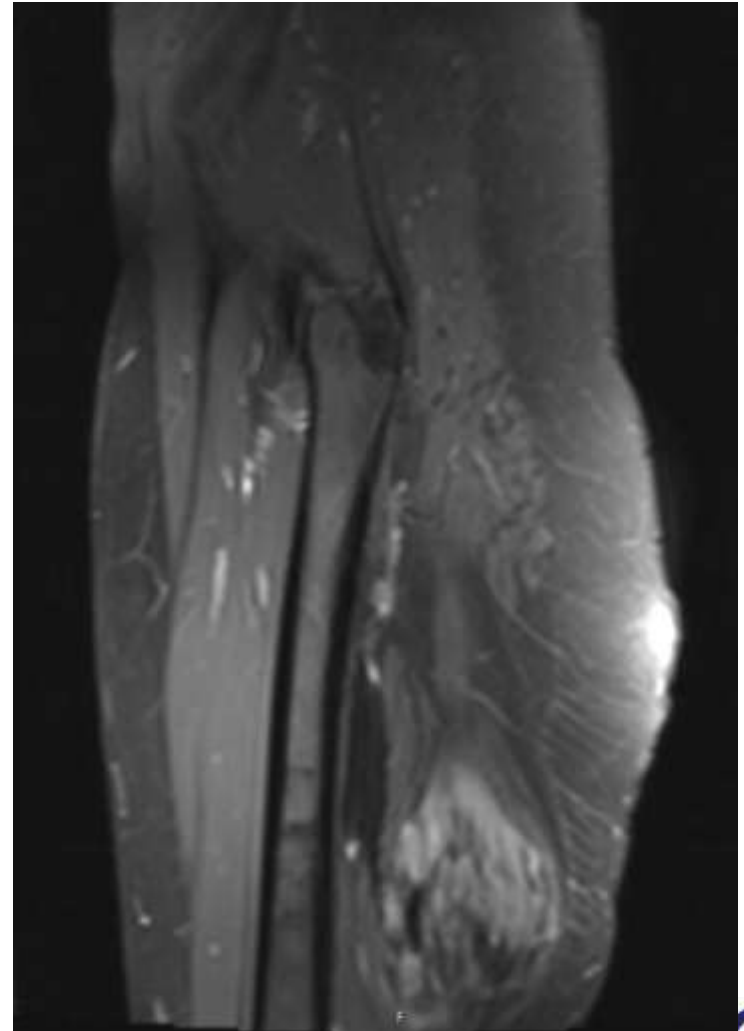
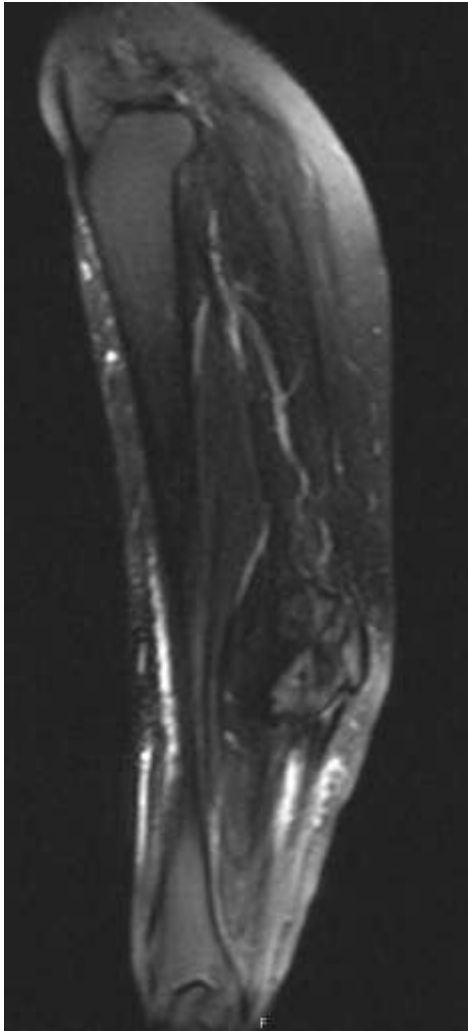
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Desmoids Vary

- Location
- Symptoms
- Appearance in images
- Appearance grossly
 - even within one tumor
- Appearance under the microscope
- Responses to treatment

Two Tumors, One Patient



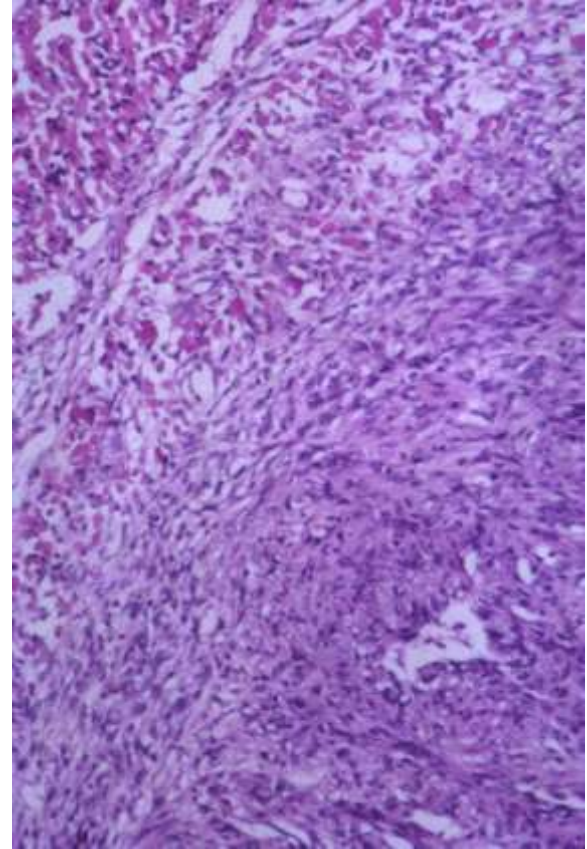
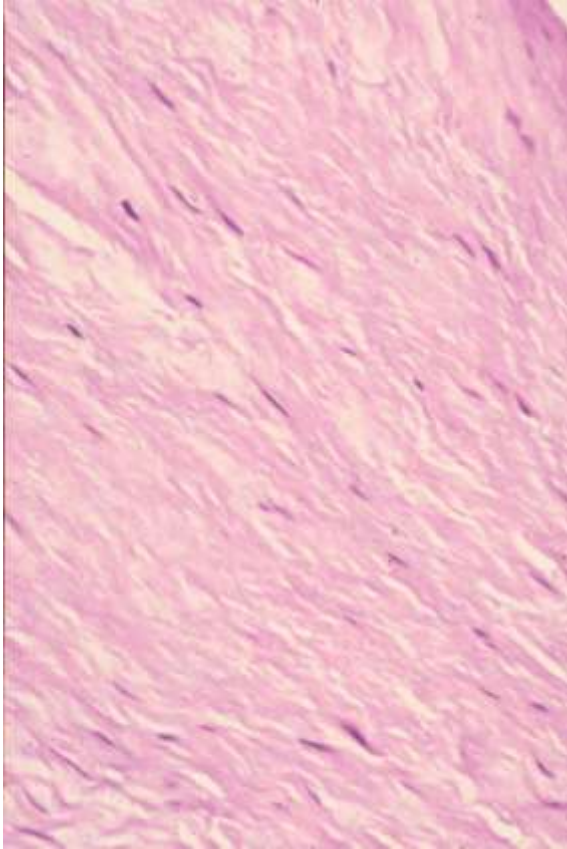
Desmoid, “Gross Specimen”



Desmoid, Fixed & Sectioned



Desmoids, Bland & “Nasty”



Principles of Treatment

- Use multidisciplinary care
 - oncologists
 - surgeons
 - pathologists
 - physical therapists
 - social workers
 - others

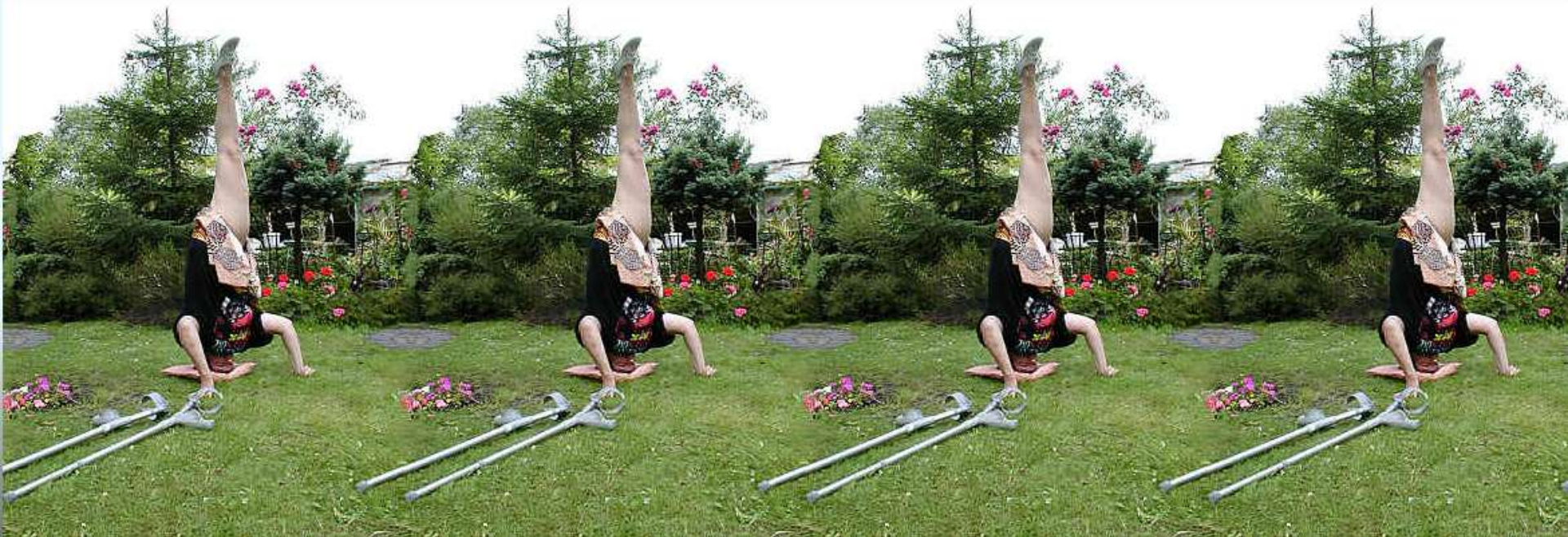
Principles of Treatment

- Use multidisciplinary care
- Avoid late effects whenever possible

We Can Do Lots of Damage

- Cyclophosphamide
 - infertility in males
 - low blood counts
 - bladder damage
- Doxorubicin
 - heart damage
- Radiation therapy
 - growth disturbances
 - malignant transformation
- Surgery
 - disfigurement
 - disability

The Dutch Hemipelvectomy Site



Principles of Treatment

- Use multidisciplinary care
- Avoid late effects whenever possible
- Minimize short-term side-effects

Short Term Side-Effects

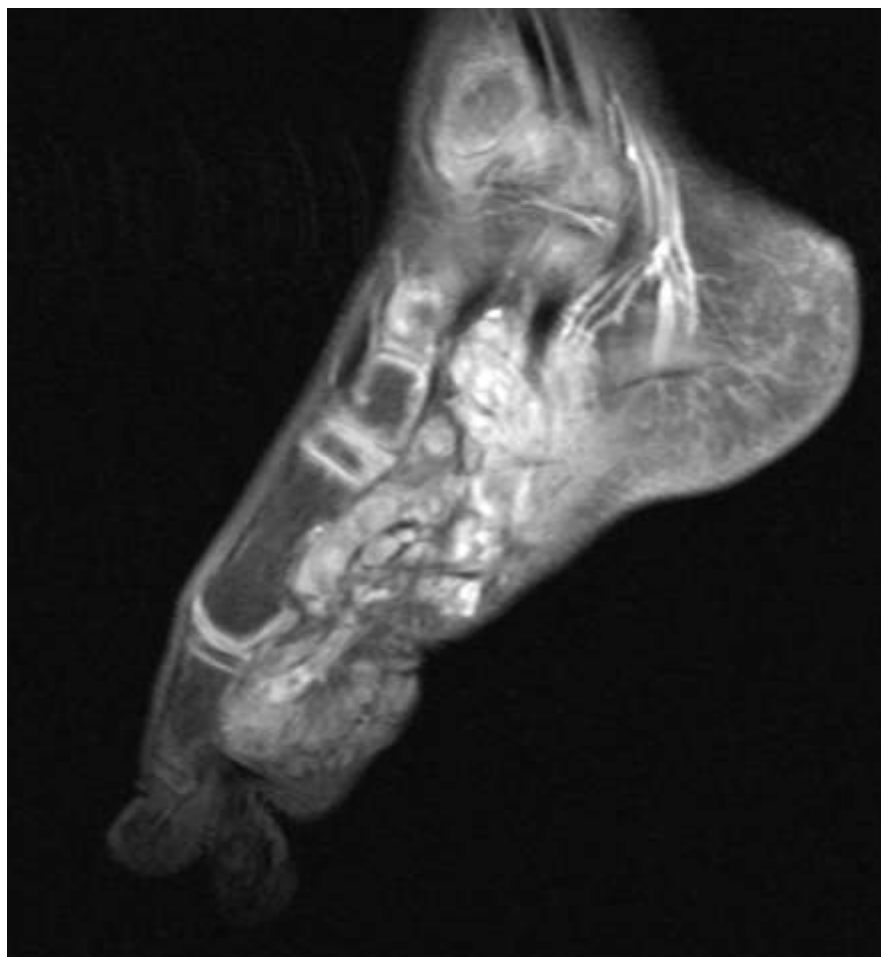
- Nausea and vomiting
- Hair loss
- Low blood counts
 - risk of infection or bleeding
 - need for transfusions
- Mouth sores
 - malnutrition
- Sensory and motor changes
 - “pins and needles”
 - clumsiness



Principles of Treatment

- Use multidisciplinary care
- Avoid late effects whenever possible
- Minimize short-term side-effects
- Eradication is not necessary

Foot Desmoid, 7 years



Principles of Treatment

- Use multidisciplinary care
- Avoid late effects whenever possible
- Minimize short-term side-effects
- Eradication is not necessary
- It's gonna take a while

Breathe... In... and... Out...

- Desmoids respond slowly
 - and may continue to shrink after treatment ends
- We cannot predict response to treatment
- They often recur or resume growing
 - and need treatment again

Speculations and Hopes

- Desmoids are a family of tumors with similar microscopic appearances – and different responses to different treatments
- Distinguishing members of the family will aid selection of treatment
- Understanding biology will lead to new treatments

