

Juvenile Xanthogranuloma (JXG)

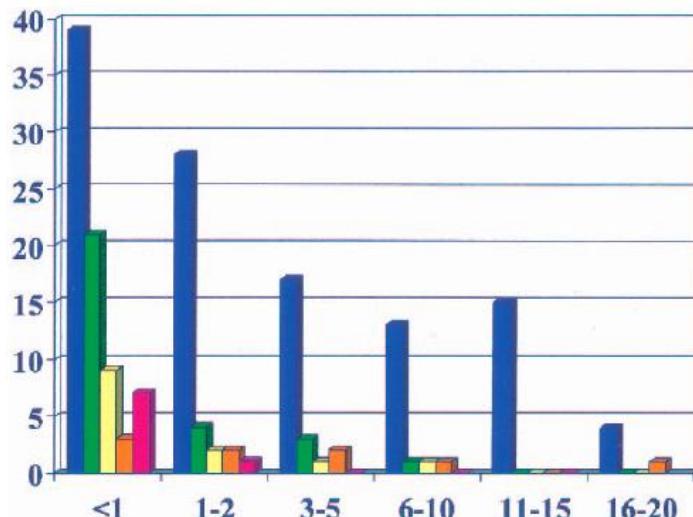
The American Journal of Surgical Pathology 2003; 27: 579-593

JXGの症状

- ・ 黄色皮膚結節 40-70% は1歳までに出現する
- ・ 通常、1 - 5 年以内に自然消退する
- ・ ごく少数例 8/174 cases (4%)

Disseminated/systemic JXG presenting in the neonatal periods.

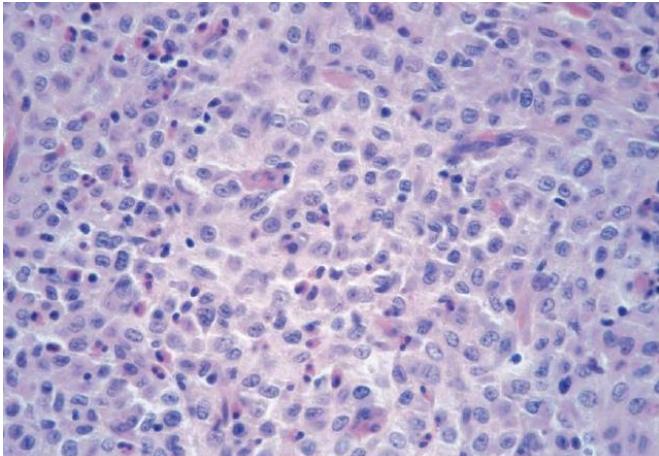
- ・ Systemic JXG の発症は、皮膚のみのJXG より低年齢 (0.3 vs 2years).
Mean 4.3 months, Median age 1 month.
- ・ ときに NF1, JMML に関連して生ずる



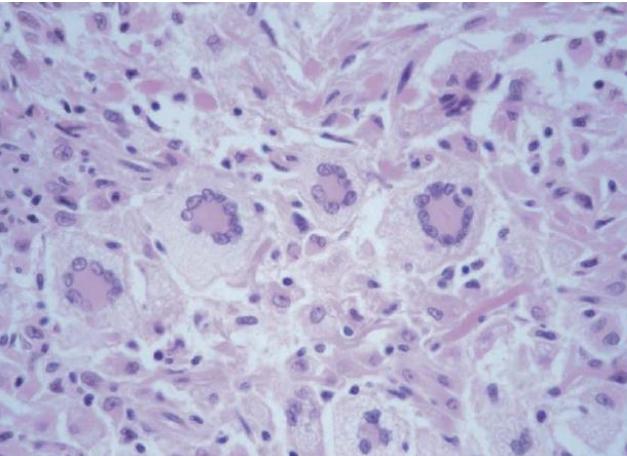
Liver, Lung, Spleen, Kidney,
Brain, Spine,
Gastrointestinal, Pancreas,
Eye/iris/orbit, Muscle,
Bone, Bone marrow, etc.

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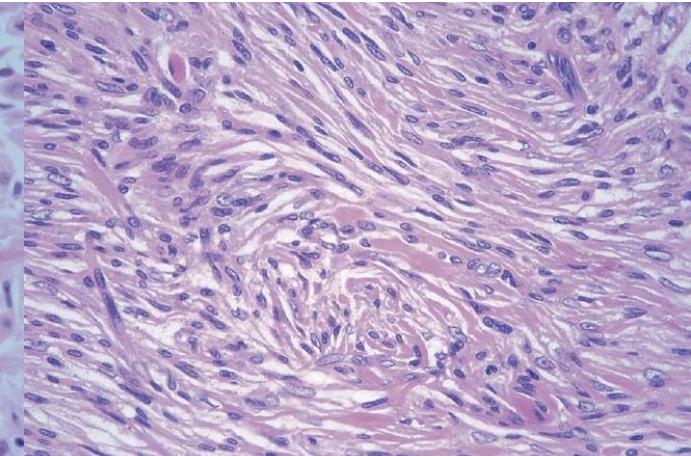
JXGの病理像



Early JXG
Monomorphic
histiocytic infiltration



Classic JXG
Foamy histiocytes
with Touton giant cell



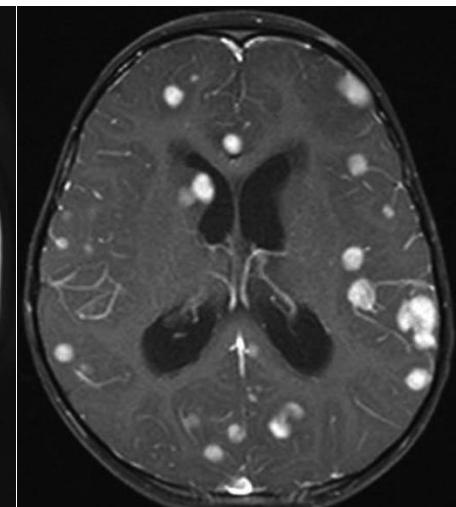
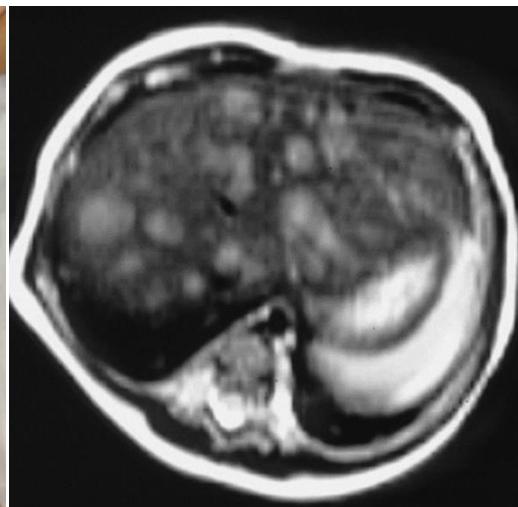
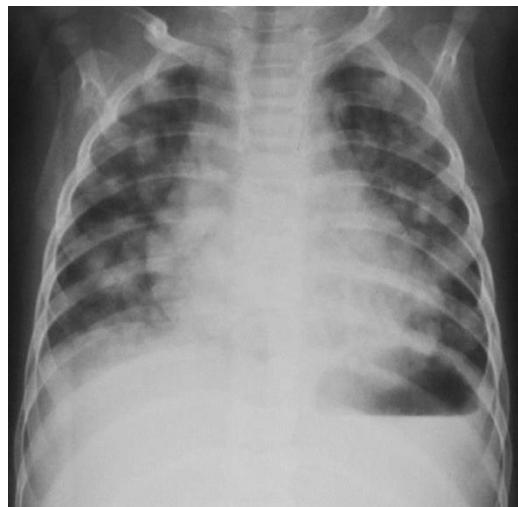
Late (transitional) JXG
Fibro-histiocytic proliferation
in a spindle cell area

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JXGの治療

- 自然軽快 ⇔ 新生児のsystemic JXGは致死的
→ LCH-based chemotherapy is a key approach
- 化学療法
 - PSL, VBL, VCR, 6MP, MTX, VP16,
 - AraC
 - Cladribine



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JXGの皮疹： 黄色の結節， Blueberry muffin rash



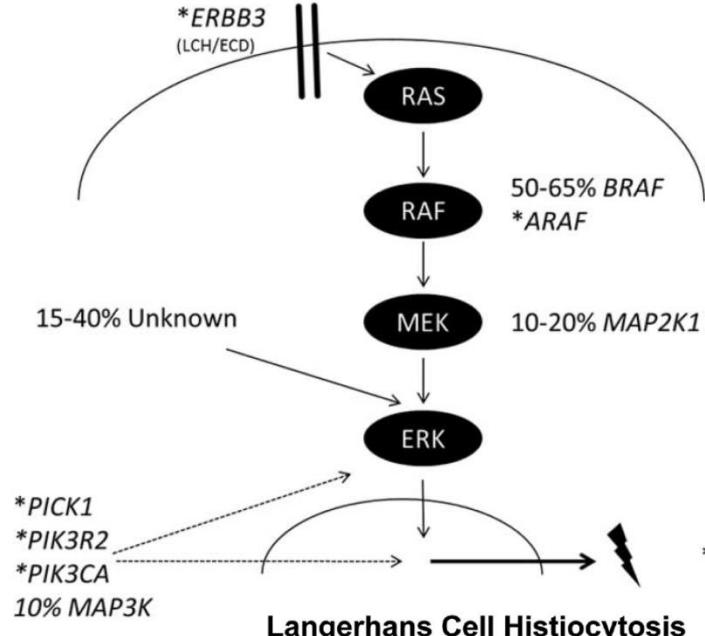
Blueberry muffin baby (散在性の紫色の丘疹, 2-6mm) :

感染（風疹、EBV、CMVなど）、溶血性疾患、悪性疾患など様々。 JXG と LCH も 鑑別疾患に挙げられる。

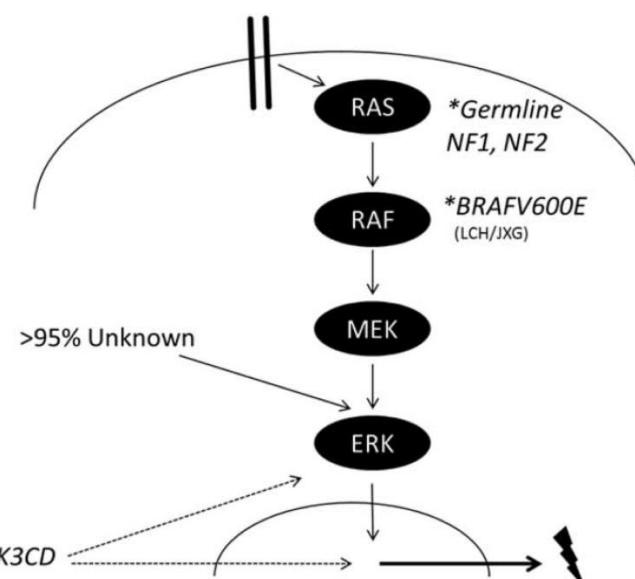
組織球症とは

“ MAPK driven myeloproliferative neoplastic disorder ”

LCH



JXG



ECD

