

ECD: Key points for Pathologists

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- 1. The classic histology of ECD is not always present: especially outside the bones
- 2. The lack of classic pathology of ECD does not exclude that diagnosis
- 3. The pathologic changes vary by site involved
- 4. None of the pathologic changes are unique to ECD

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ECD HISTOLOGIC FINDINGS

Foamy histiocytes
Touton giant cells
Non-foamy histiocytes
Fibrosis
Lymphocytes
Plasma cells

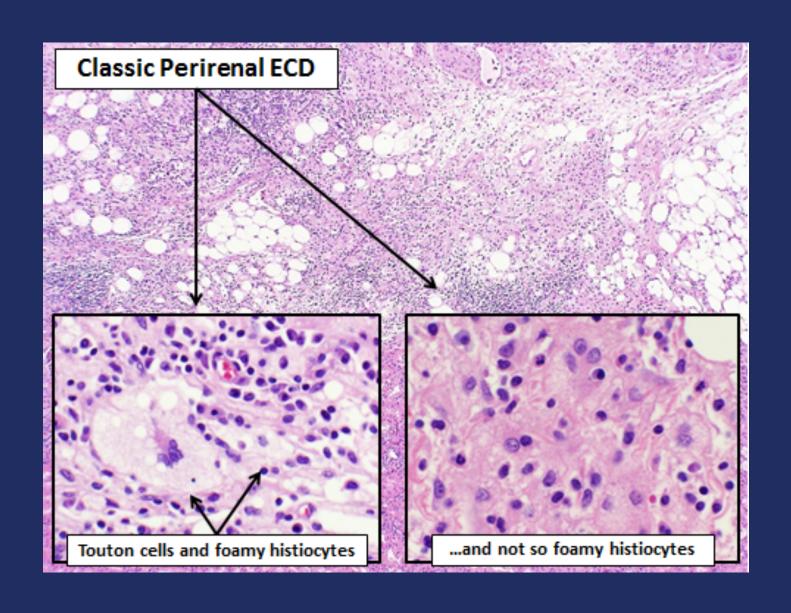
None specific/unique

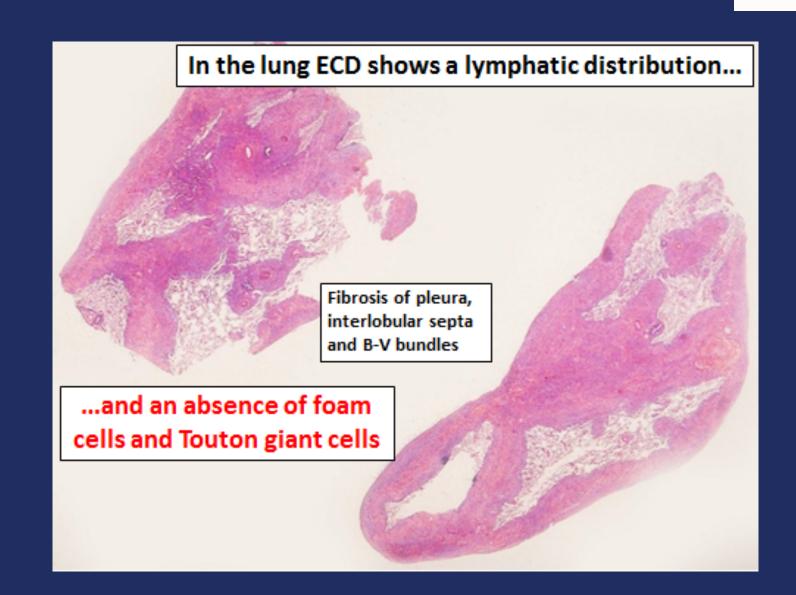
The %'s vary

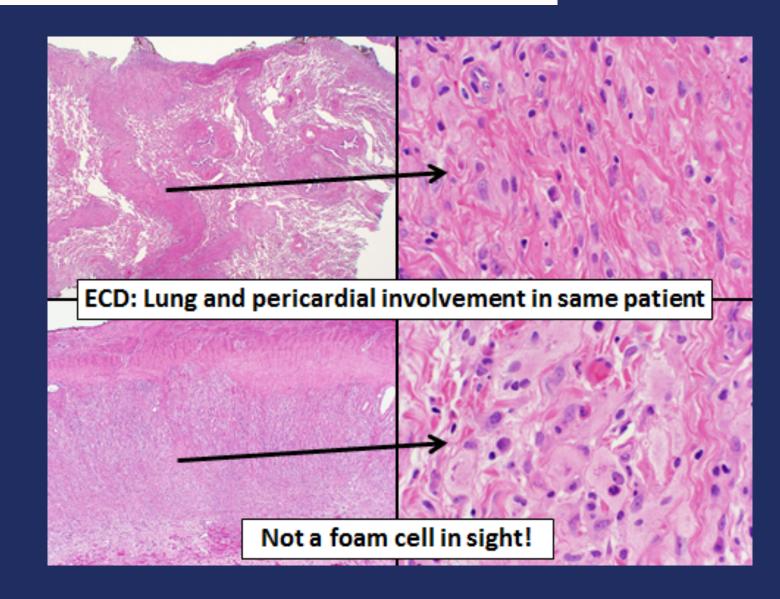
None universally present

Histiocytes are:

CD68+ CD163+ Fact13a+ s-100+/- CD1aBRAFV600e staining +/- (but little data yet)







Unpublished Data from MSK

(Courtesy A Dogen, E Diamond, WD Travis et al.)

Pathologic findings in 43 patients with ECD at a variety of sites, some with multiple biopsies

Foamy macrophages: ~55%
Touton giant cells: ~22%

Fibrosis: 100%

CD68/CD163⊕: 100%

ECD RE-ASSESSMENT

Identification of BRAF V600E (and other) mutations

Up to 100% ECD patients (specificity unknown)
~50% or less of pulmonary LCH
Found in many other tumors (melanoma, PTC, colon, NSCLCa)

Re-assessment of pathogenesis; options for targeted therapies.

Oncology and hematopathology should lay claim to ECD in light of recent data suggesting neoplastic histiocytes