

Tumors of Fibrous Tissue Involving the Skin

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Immunohistochemical stains helpful in distinguishing:

Stain	DF	DFSP
CD34		
Factor 13a		
bcl-2		
Tenascin		
CD44		

Answers

Stain	DF	DFSP
CD34		+
Factor 13a	+	
bcl-2		+
Tenascin	+ (at DEJ)	
CD44	+	

Risk of Metastases?

Disease	Low Risk	High Risk
Atypical fibroxanthoma		
Epithelioid sarcoma		
Cellular dermatofibroma		
DFSP		
Malignant fibrous histiocytoma		

Answers

Disease	Low Risk	High Risk
Atypical fibroxanthoma	+	
Epithelioid sarcoma		+
Cellular dermatofibroma	+	
DFSP	+	
Malignant fibrous histiocytoma		+

Pertinent histopathological differential diagnostic considerations for epithelioid sarcoma include:

- Basal cell carcinoma, nodular type
- Malignant extrarenal rhabdoid tumor
- Granuloma annulare
- Amelanotic melanoma
- Epithelioma of Bordst-Jadosshn

May have more than one answer.

Answers

- Malignant extrarenal rhabdoid tumor
- Granuloma annulare
- Amelanotic melanoma

Key Associations:

Disease	Histopathology
PNET	t(X,18)(p11;q11)
Infantile Digital Fibromatosis	Birt-Hogg-Dube syndrome
Trichodiscoma	Eosinophilic cytoplasmic inclusion bodies
Synovial sarcoma	MIC2 gene overexpression
Sclerotic fibroma	Eosinophilic cytoplasmic inclusion bodies

Answers

Disease	Histopathology
PNET	MIC2 gene overexpression
Infantile Digital Fibromatosis	Eosinophilic cytoplasmic inclusion bodies
Trichodiscoma	Birt-Hogg-Dube syndrome
Synovial sarcoma	t(X,18)(p11;q11)
Sclerotic fibroma	Cowden's syndrome