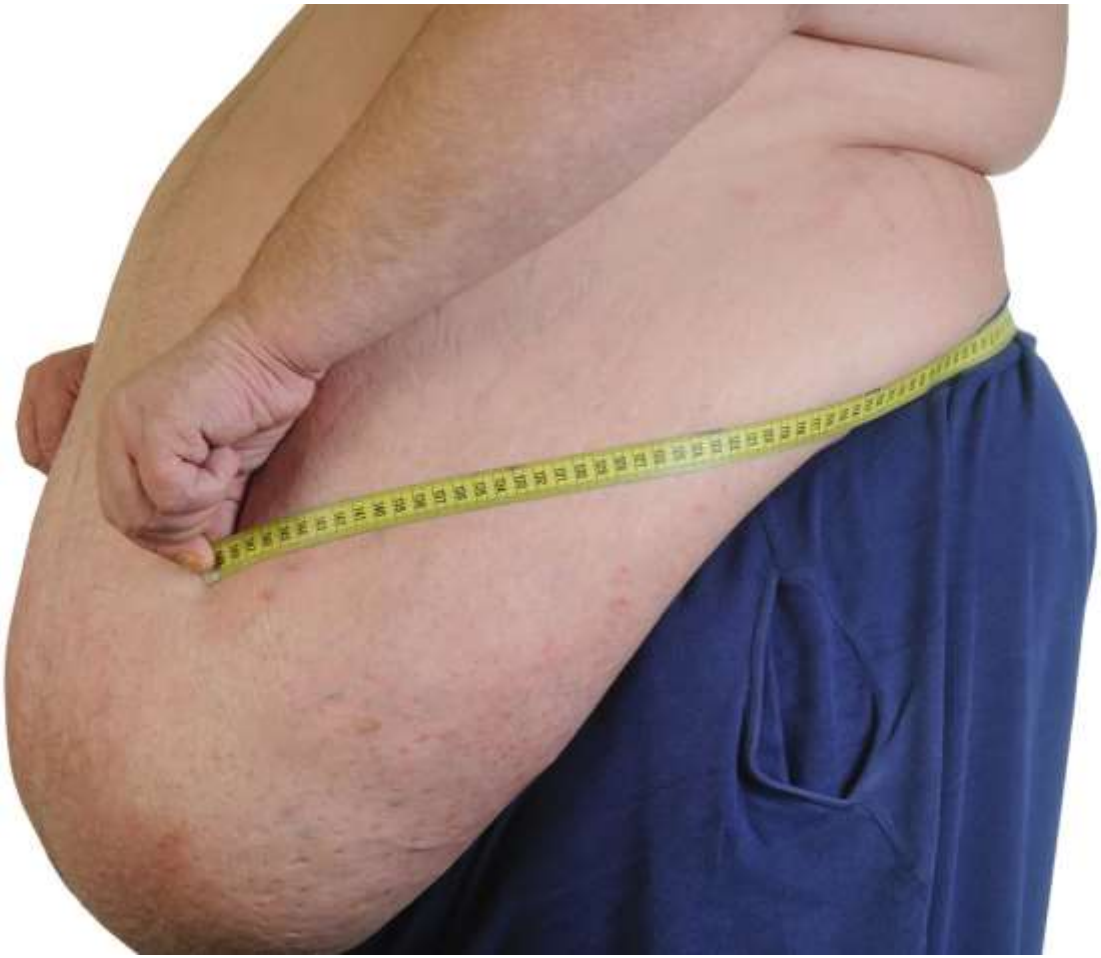
Mestcellen

dr. Herman Bril

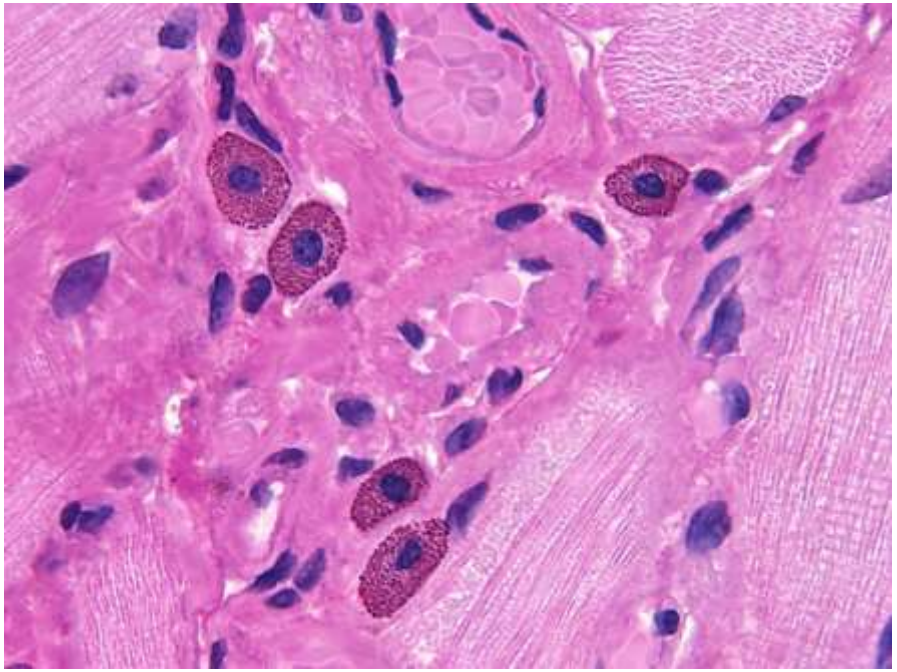
P. Ehrlich 1879: Mestcellen

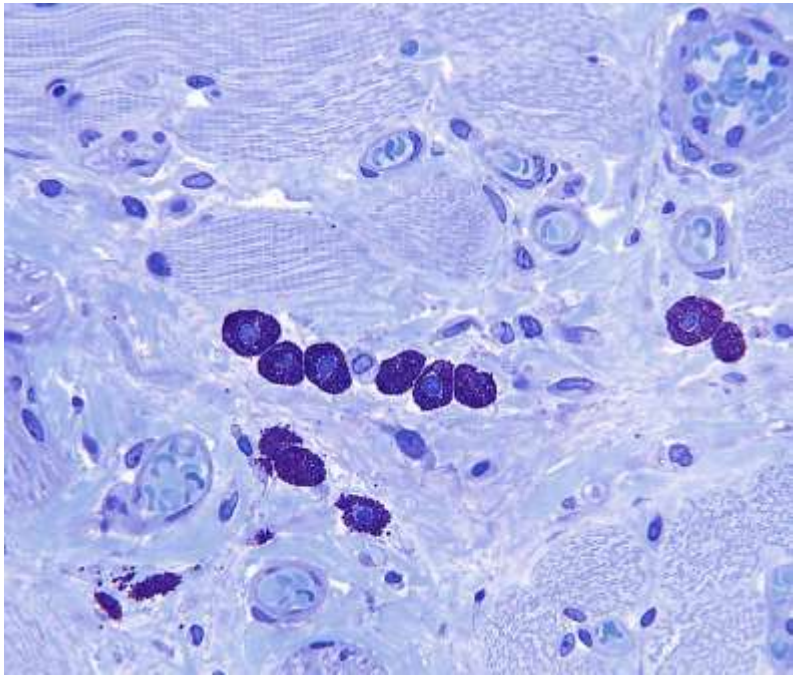
("vetgemeste" overvoede grote bindweefsel cellen vol met korrels)

Vetgemest

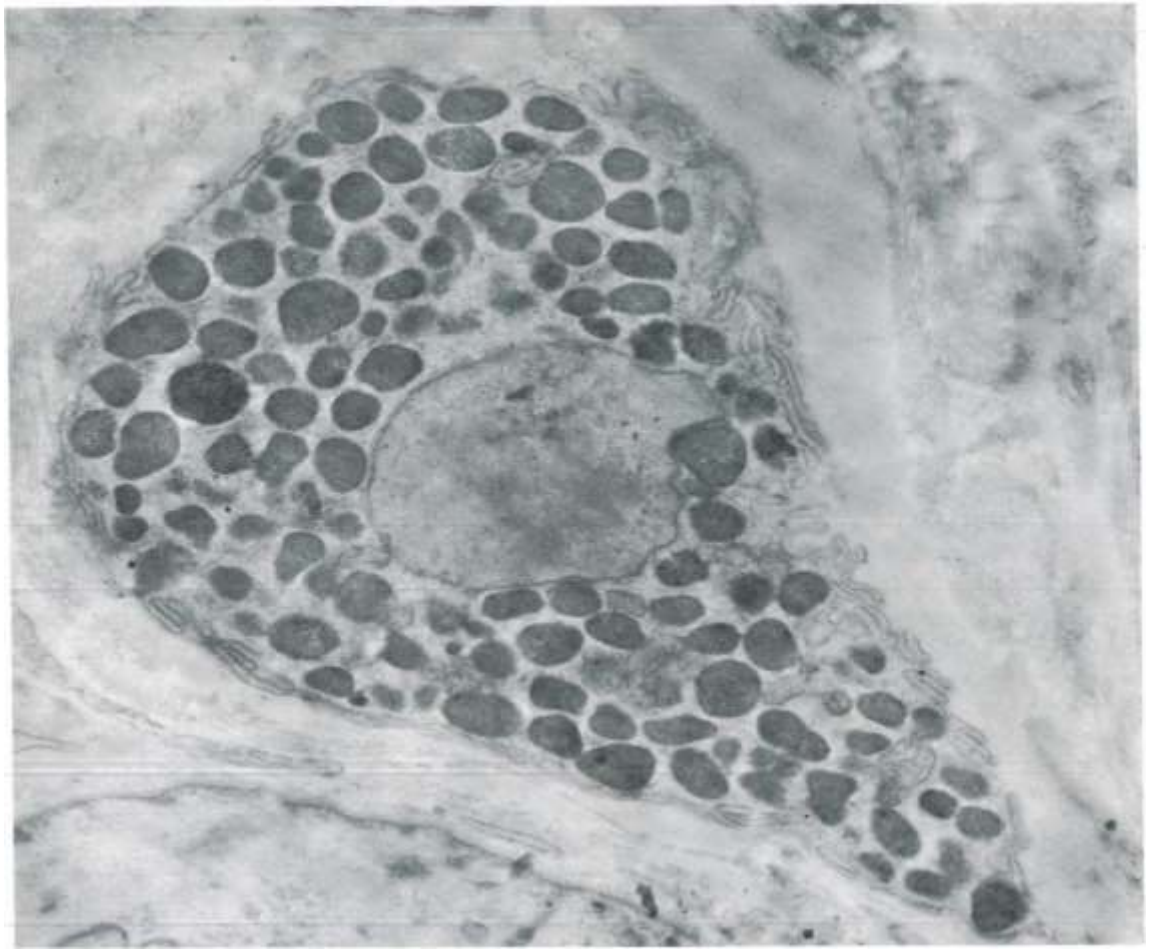


Histologie

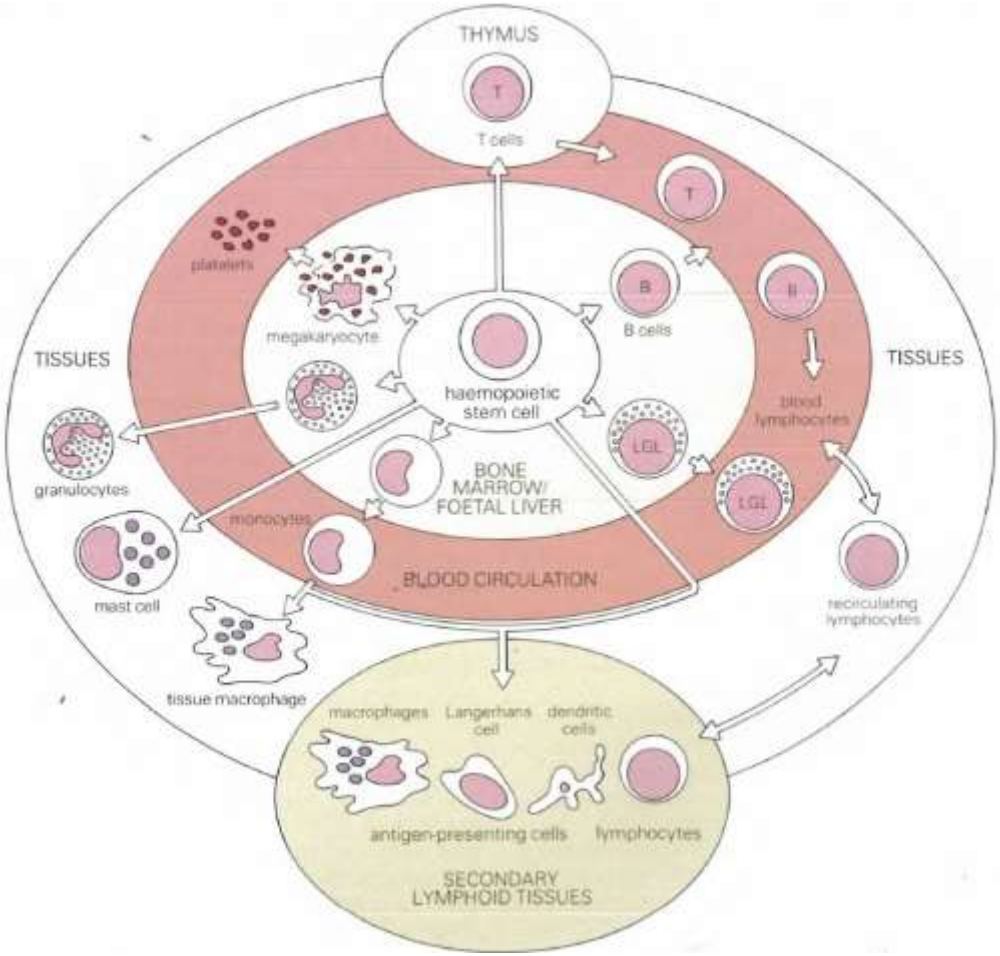


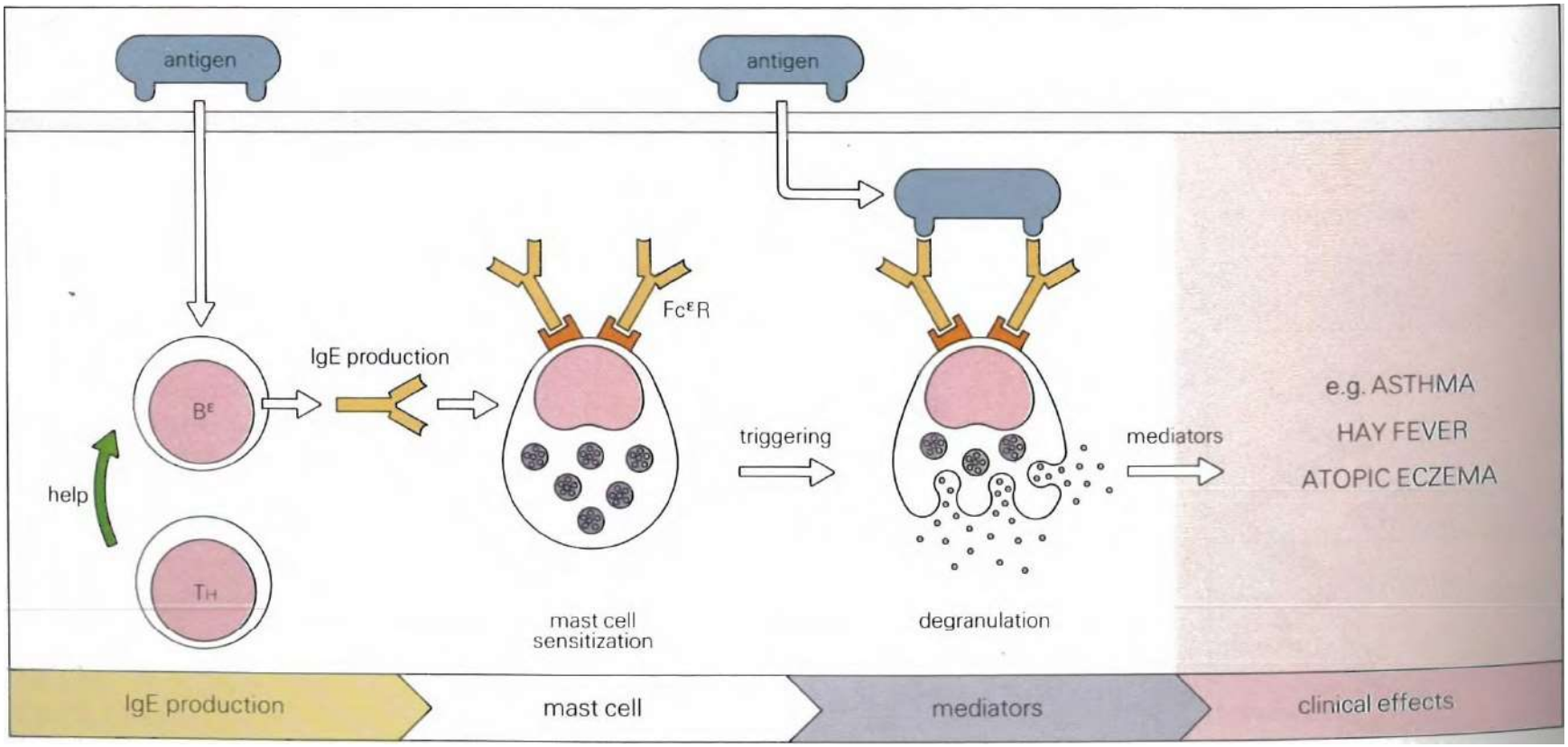


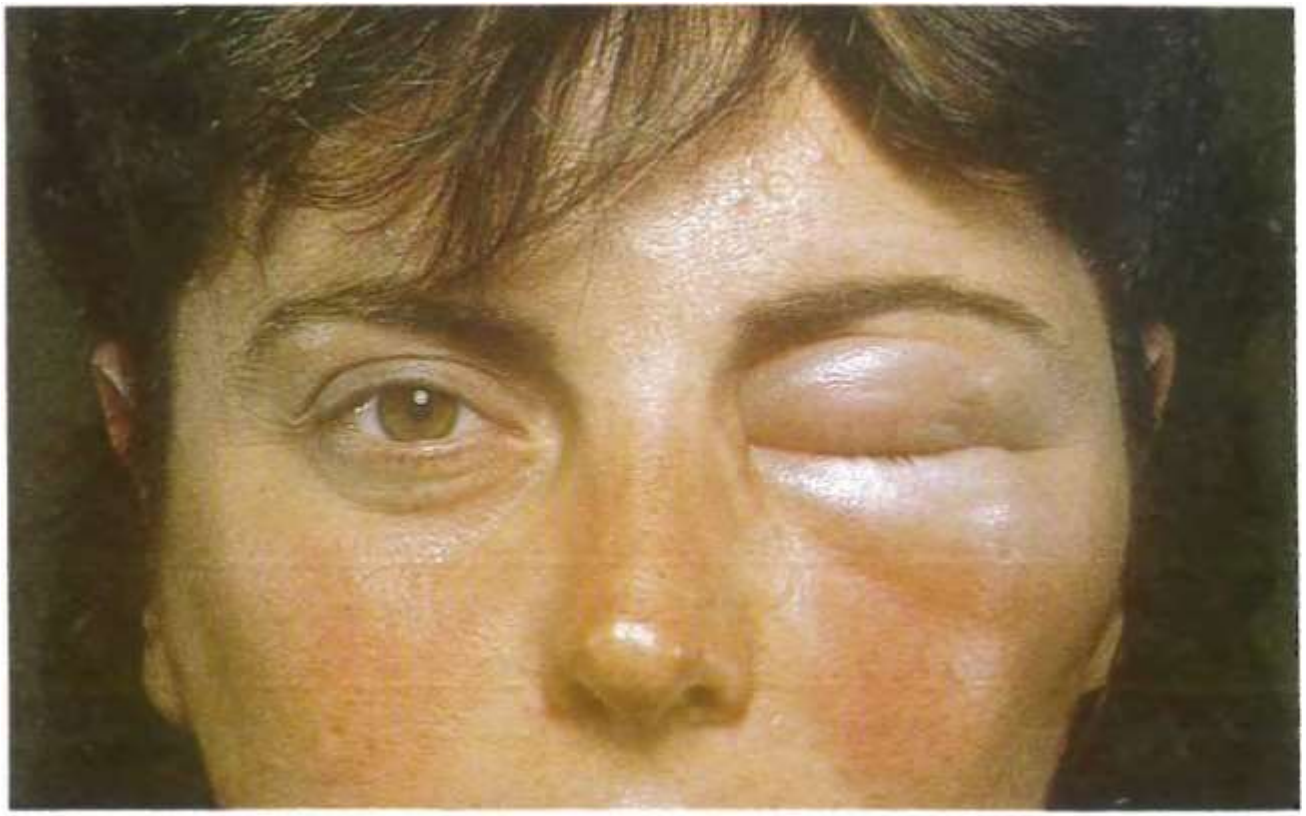
Mestcellen

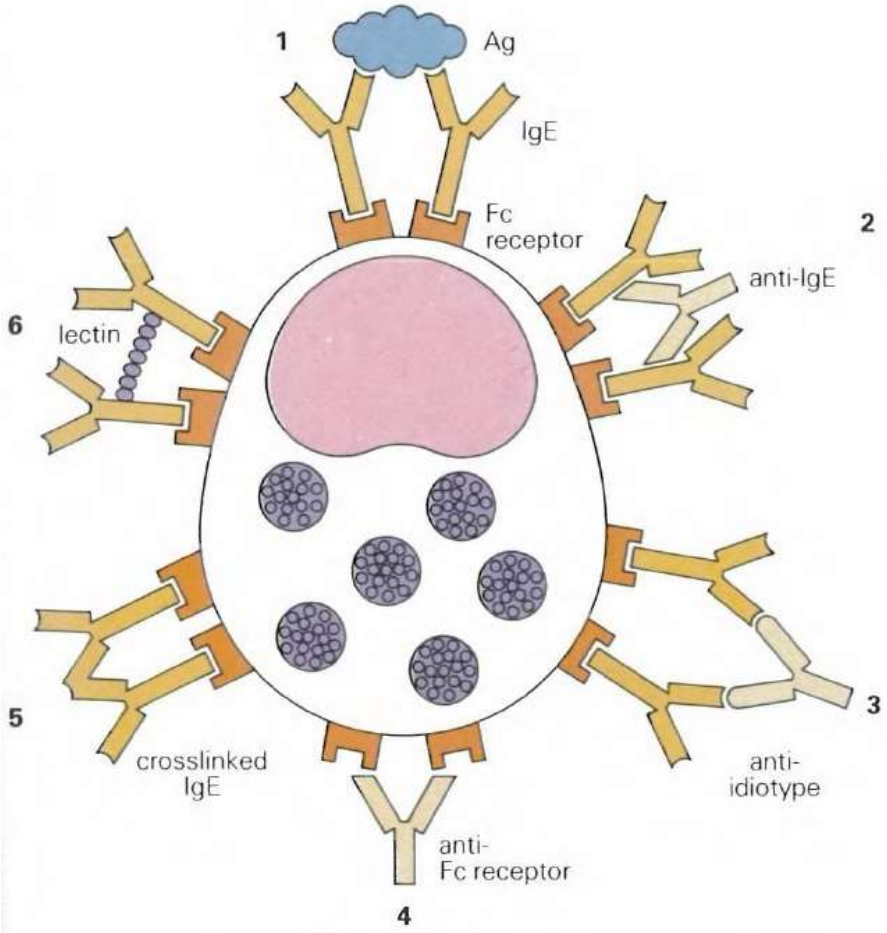


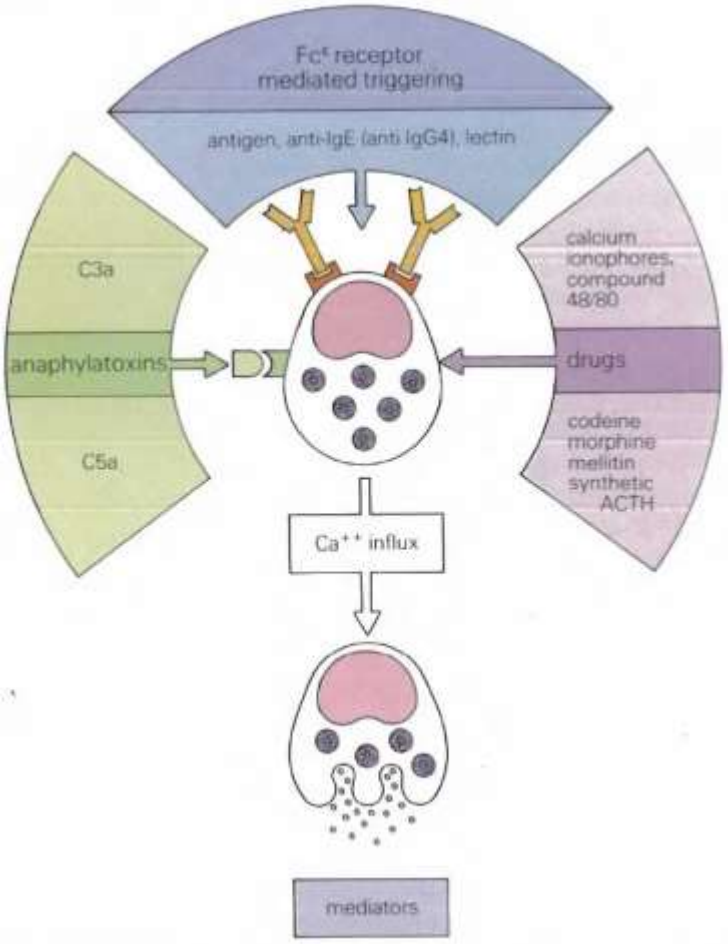
Oorsprong


THYMUS
T
T cells
T
platelets
megakaryocyte
B
B cells
B
TISSUES
TISSUES
haemopoietic stem cell
blood lymphocytes
LGL
granulocytes
BONE MARROW/ FOETAL LIVER
LGL
monocytes
BLOOD CIRCULATION
mast cell
recirculating lymphocytes
tissue macrophage
macrophages
Langerhans cell
dendritic cells
antigen-presenting cells
lymphocytes
SECONDARY LYMPHOID TISSUES

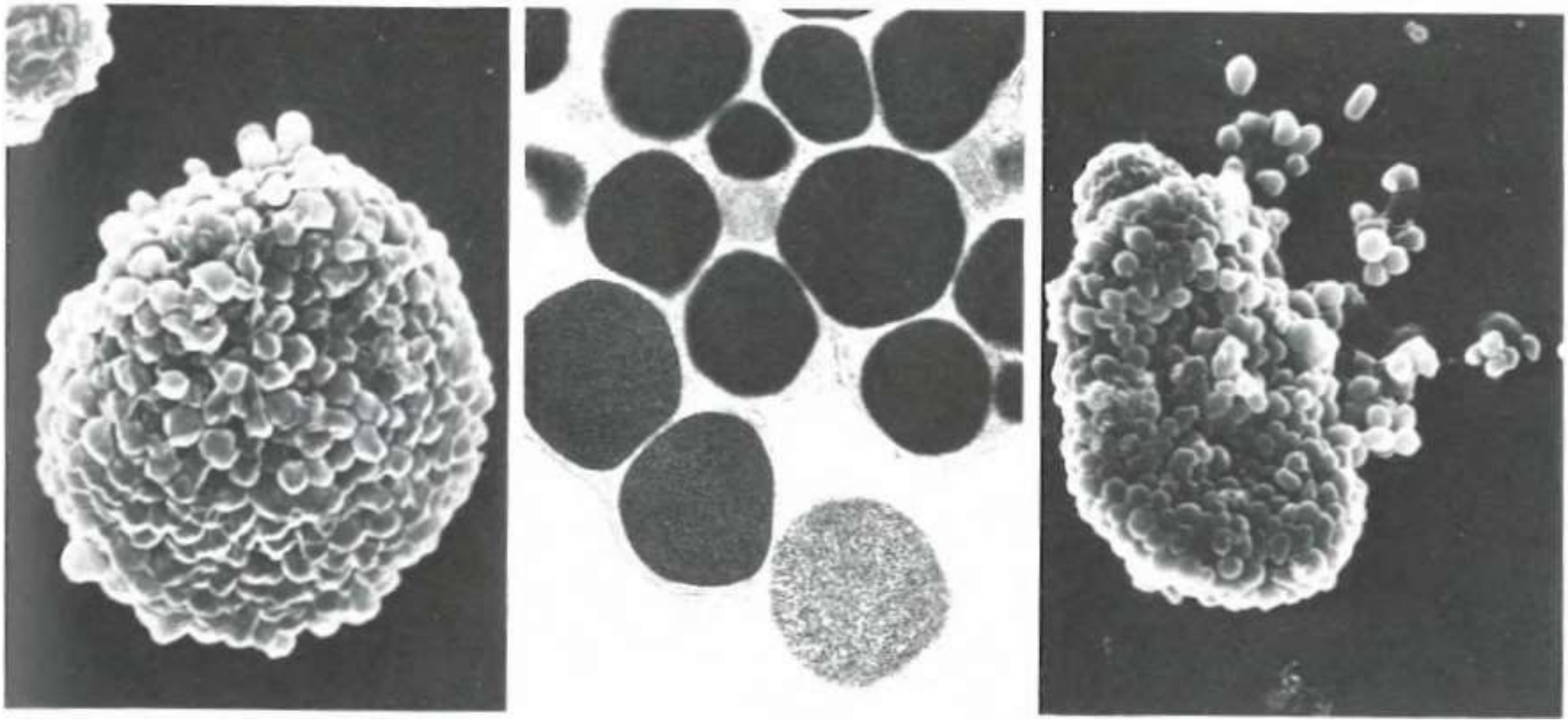

antigen
antigen
IgE production
help
triggering
mediators
e.g. ASTHMA
HAY FEVER
ATOPIC ECZEMA
mast cell
sensitization
degranulation
IgE production
mast cell
mediators
clinical effects




1
Ag
IgE
Fc
receptor
2
anti-IgE
6
lectin
5
crosslinked
IgE
3
anti-
idiotype
anti-
Fc receptor
4


Fc receptor
mediated triggering
antigen, anti-IgE (anti IgG4), lectin
C3a
anaphylatoxins
C5a
calcium
ionophores,
compound
48/80
drugs
codeine
morphine
mellitin
synthetic
ACTH
Ca++ influx
mediators

granule-associated preformed mediators		
histamine	Mol.Wt. = 111	vasodilation, increased capillary permeability chemokinesis bronchoconstriction
heparin	Mol.Wt. = 60,000	anticoagulant
enzymes	tryptase (Mol.Wt. = 130,000) β-glucosaminidase (Mol.Wt. = 150,000)	proteolytic C3 convertase cleaves glucosamine residues
chemotactic and activating factors	ECF-A (Mol.Wt. = 380/2000) NCF (Mol.Wt. >750,000) PAF (Mol.Wt. =600)	chemotaxis of eosinophils, neutrophils platelet activation
newly formed mediators		
SRS (LTC_4+LTD_4) chemotactic leukotrienes (LTB_4)	lipoxygenase pathway products	vasoactive, broncho-constriction, chemotactic and/or chemokinetic
prostaglandins thromboxanes	cyclooxygenase products	bronchial muscle contraction, platelet aggregation, vasodilation



Symptomen veroorzaakt door mediatoren

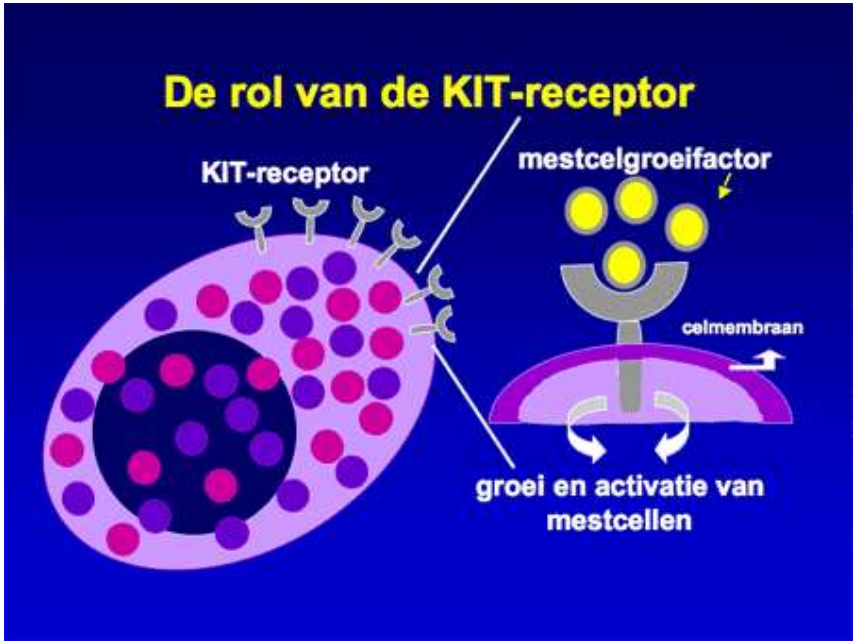
- Opvliegers met een rode kleur, hartkloppingen, “warm worden”
- Buikpijn met diarree
- Spierpijn
- Botpijn met botontkalking
- Aanvallen van onwel worden, soms met lage bloeddruk, bewustzijnsverlies (“anafylactische shock”)

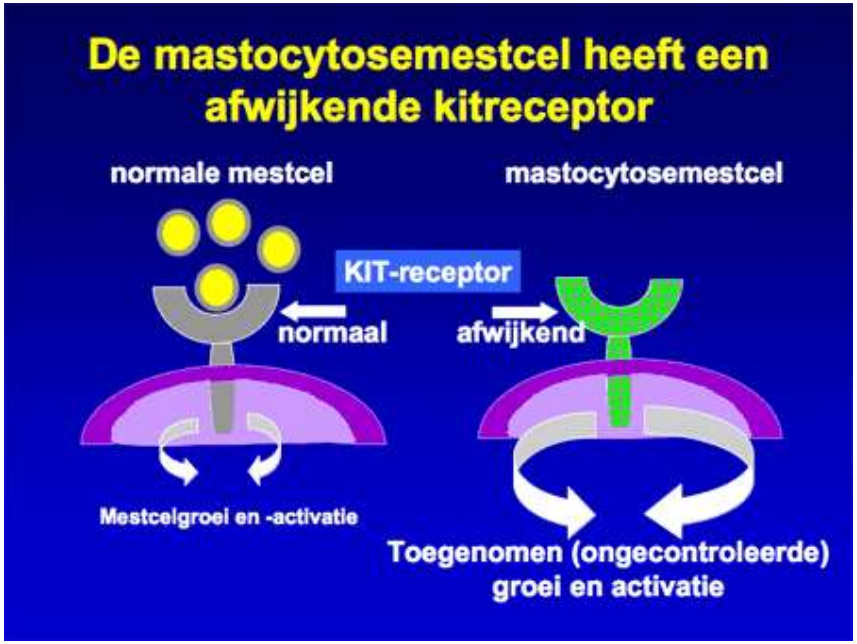
Diagnostiek

- Urine tryptase (afbraakproduct van histamine)
- Huidbiopt
- Beenmergonderzoek

Therapie

- Gericht op het “kalmeren” van de mestcellen en het wegvangen van vrijgekomen stoffen (“symptoombestrijding”) (histamine blokkers)
- Bij kwaadaardige vormen: celdodende middelen
- Epi-pen

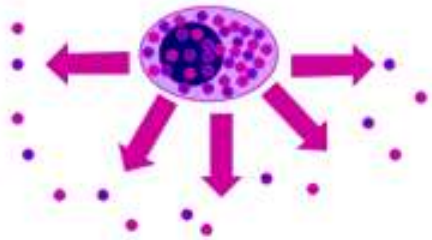

De rol van de KIT-receptor
KIT-receptor
mestcelgroeifactor
celmembraan
groei en activatie van mestcellen


De mastocytosemestcel heeft een afwijkende kitreceptor
normale mestcel
mastocytosemestcel
KIT-receptor
normaal
afwijkend
Mestcelgroei en -activatie
Toegenomen (ongecontroleerde) groei en activatie

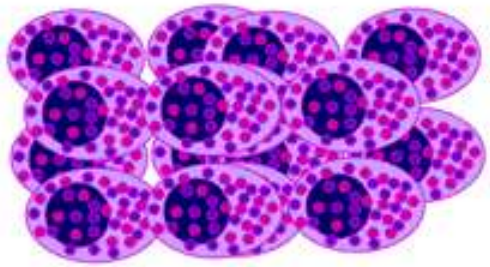
Mastocytose: toename van afwijkende mestcellen in huid, beenmerg, darmstelsel, milt, lever of botten

Dit kan resulteren in de volgende problemen

Plaatselijke of algemene symptomen door uitscheiding van mestcelfactoren

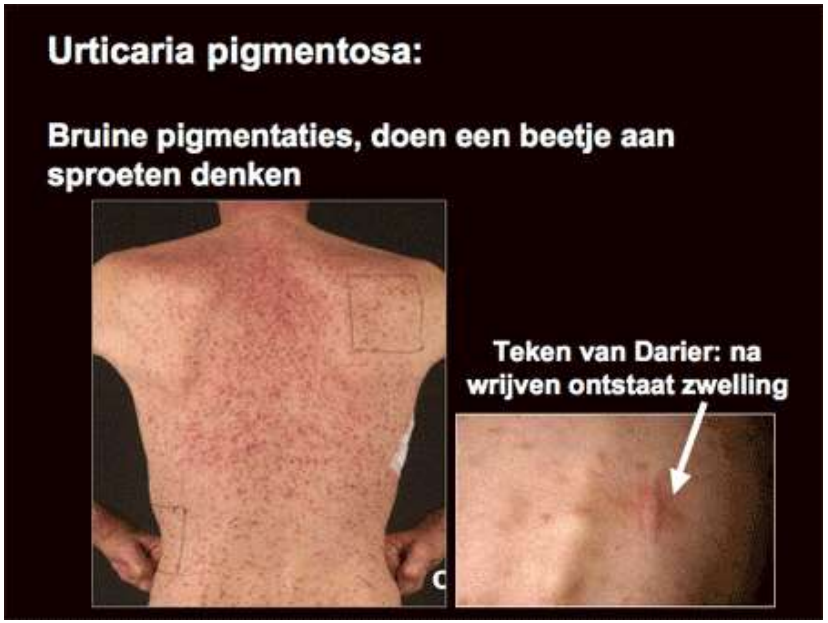


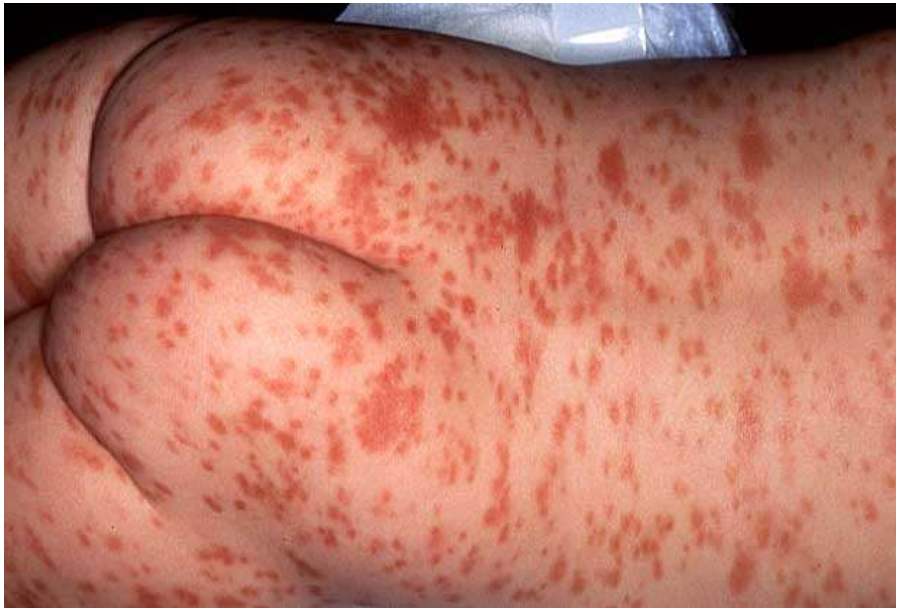
Toename van mestcellen in beenmerg en andere weefsels



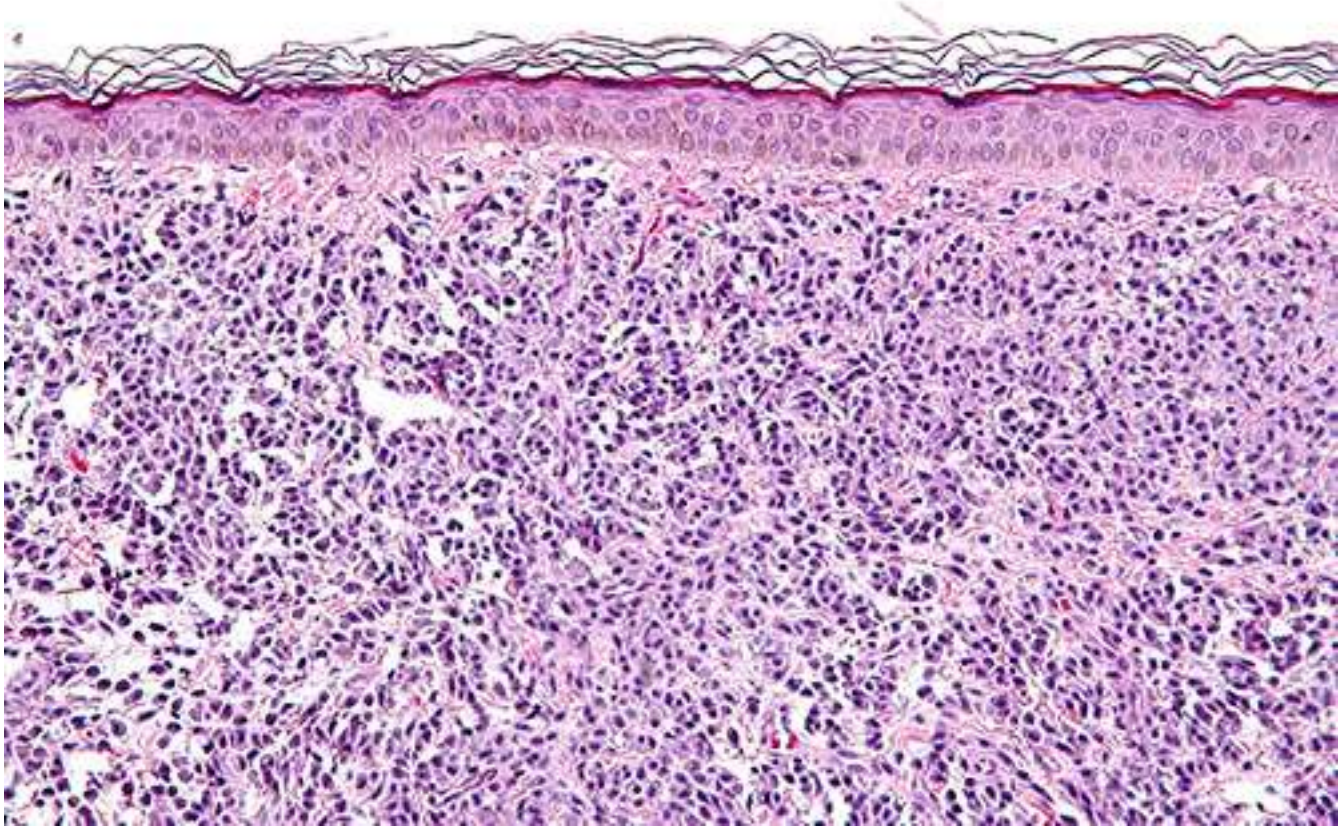
Indeling mastocytose volgens de WHO-classificatie	
Alleen huidmastocytose	Het meest voorkomend
Indolente systemische mastocytose	Het meest voorkomend
Systemische mastocytose met bijkomende andere bloedziekte	Zeldzaam
Agressieve systemische mastocytose	Zeldzaam
Overige vormen (mestcelleukemie, mestceltumoren)	Zeer zeldzaam
WHO = World Health Organization	

Huidafwijkingen

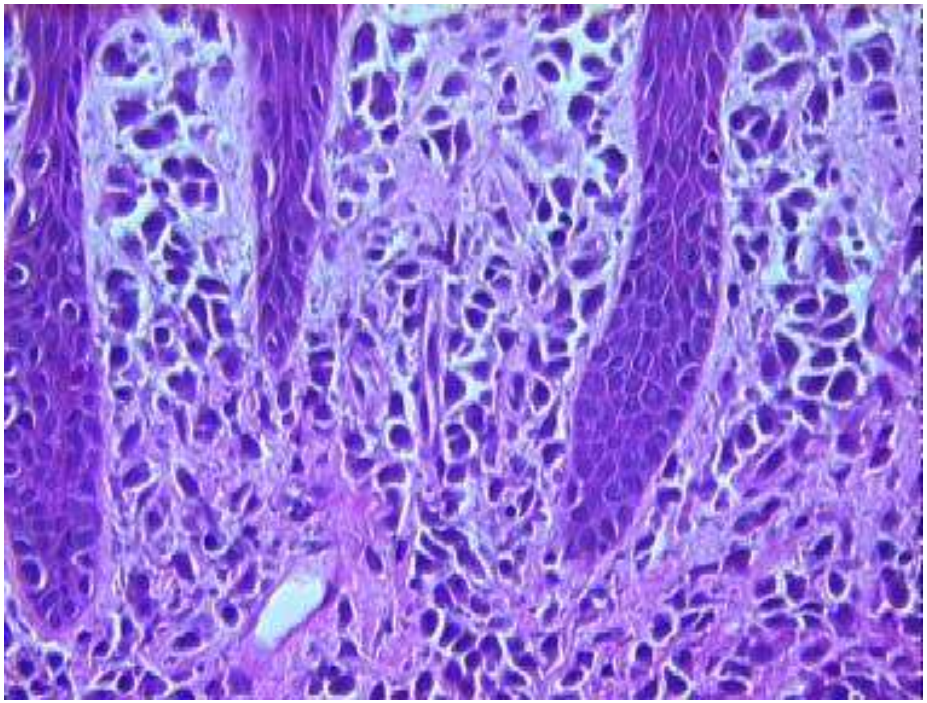




Huidbiopt

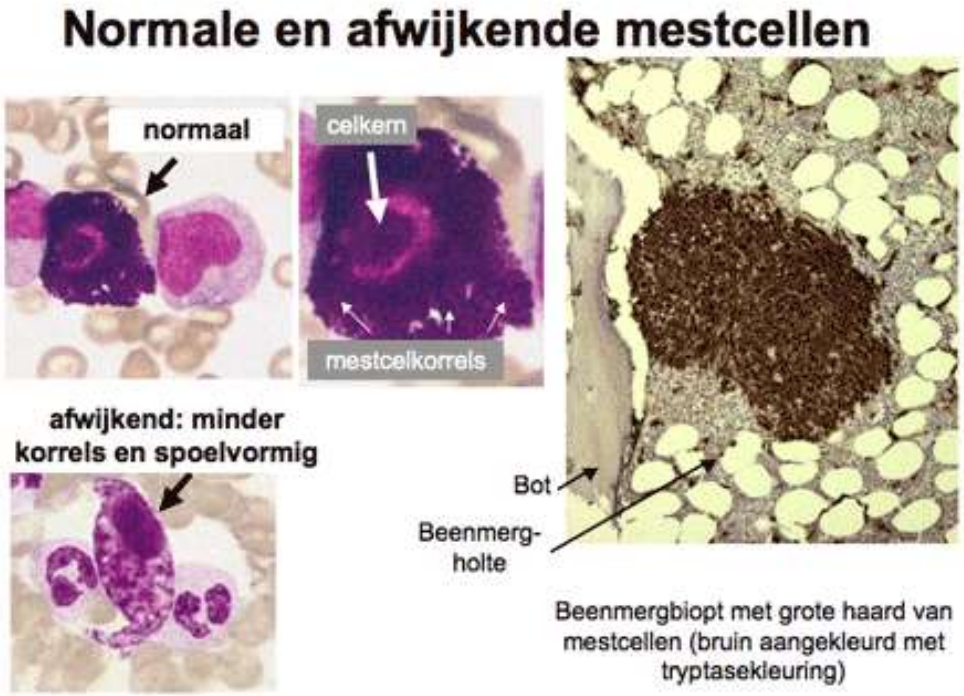


Huidbiopt grotere vergroting

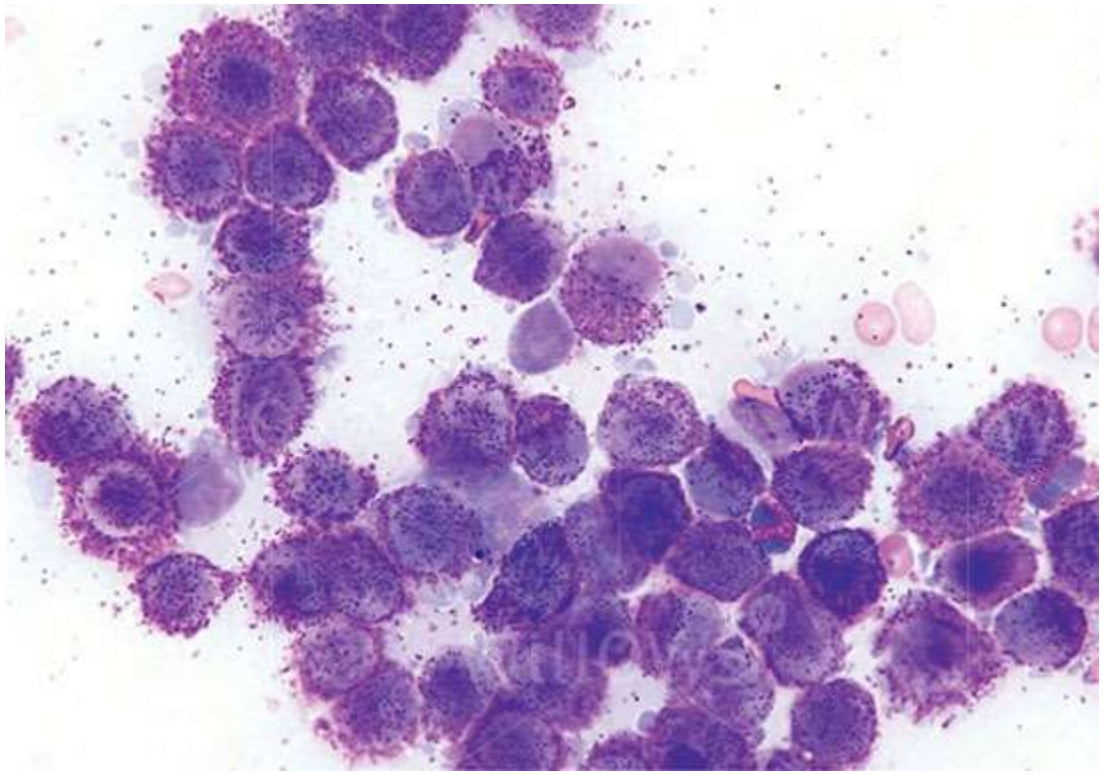


CD117

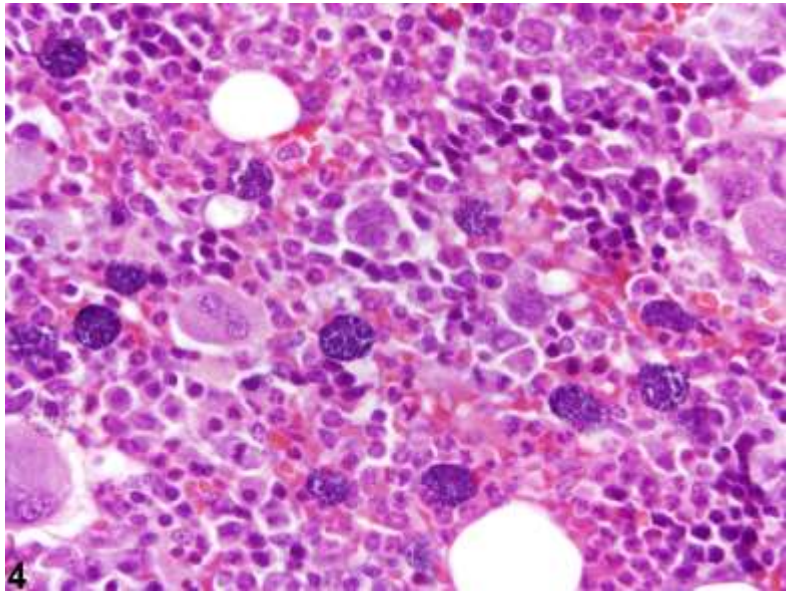
Systemische mastocytose



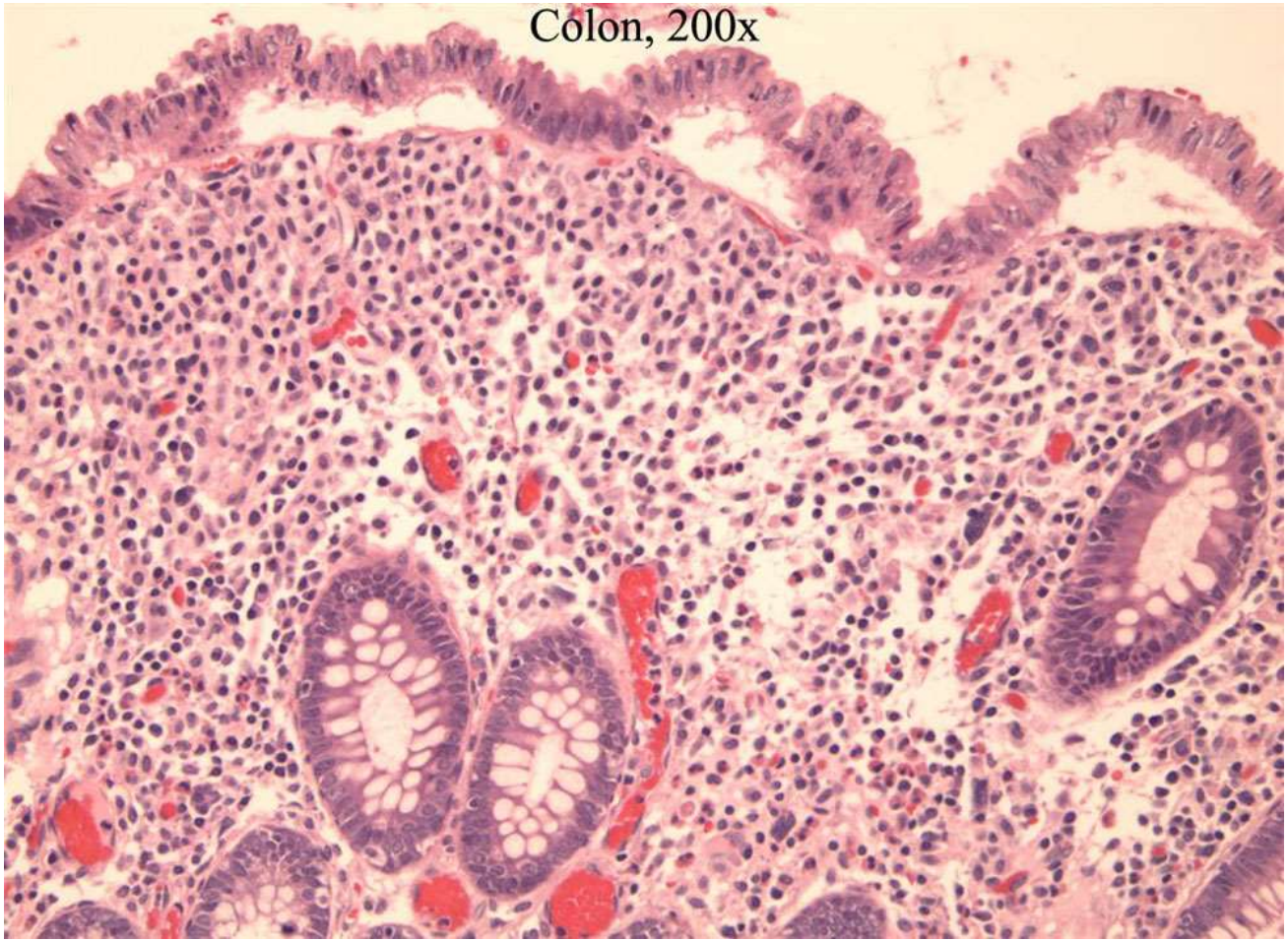
Beenmerg aspiraat



Beenmerg histologie



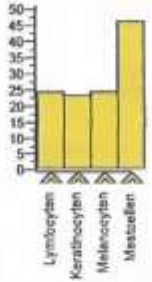
Mastocytose in de darm



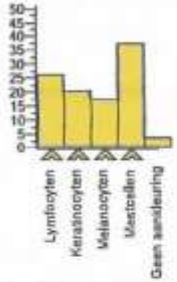
Giemsa

Giemsa aankleuring

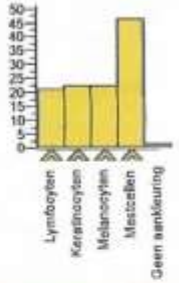
2014.1 A



2014.1 B



2014.1 C

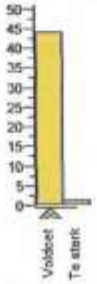


Legenda

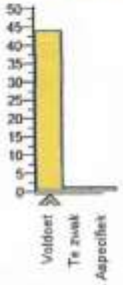
Giemsa

Giemsa kleuring voldoet

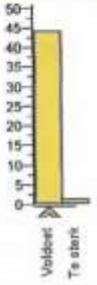
2014.1 A



2014.1 B

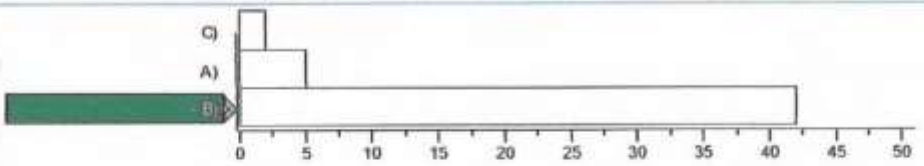


2014.1 C

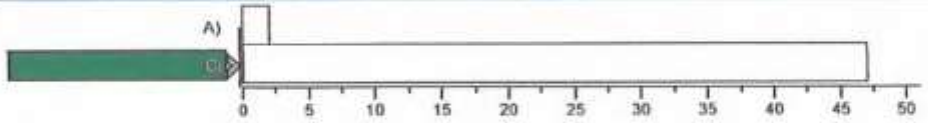


Conclusievragen

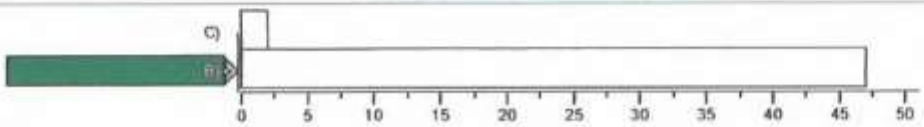
Monster : 2014.1 A	Score
Wat is uw eindconclusie voor Huid I A) Huid zonder afwijkingen B) Huid met mestcelafwijking C) Huid met afwijking, vervolgonderzoek m.b.v. Immunohistochemie geadviseerd D) Niet te beoordelen vanwege kwaliteit coupe/kleuring	2

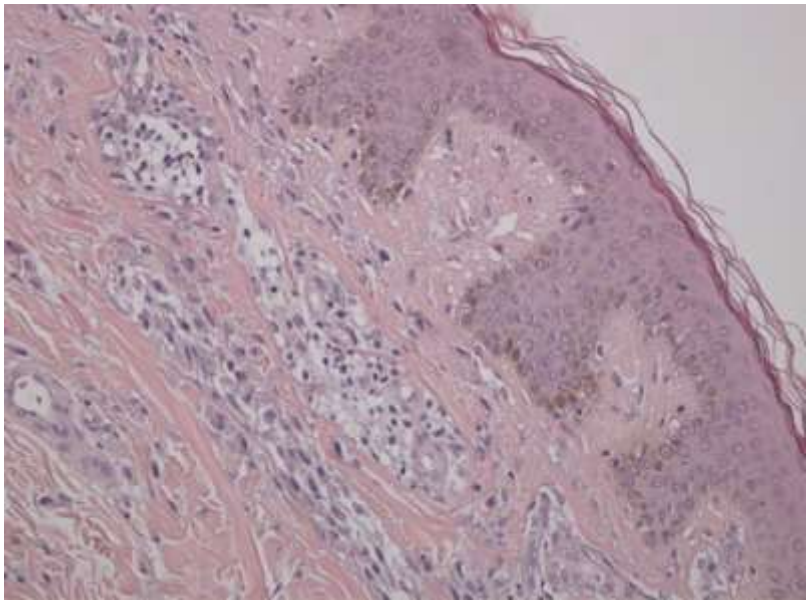


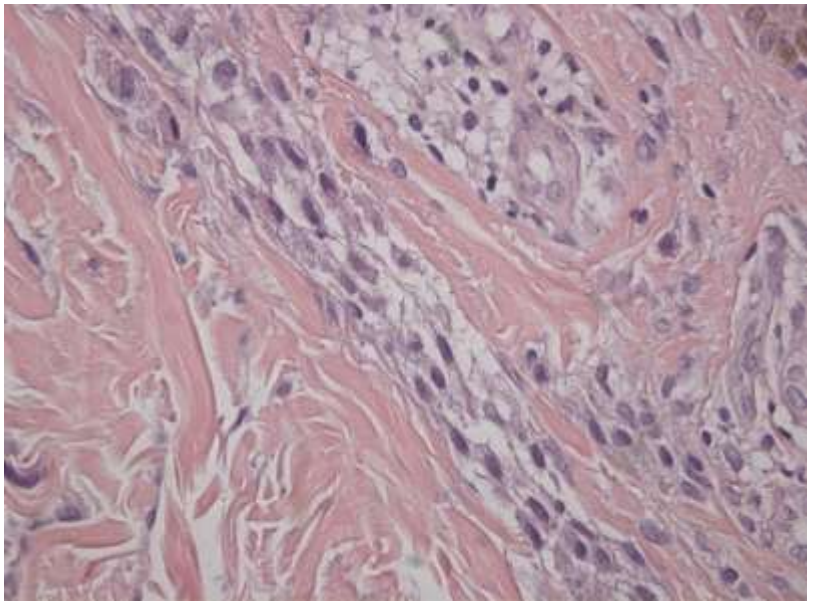
Monster : 2014.1 B	Score
Wat is uw eindconclusie voor Huid II A) Huid zonder afwijkingen B) Huid met mestcelafwijking C) Huid met afwijking, vervolgonderzoek m.b.v. Immunohistochemie geadviseerd D) Niet te beoordelen vanwege kwaliteit coupe/kleuring	2

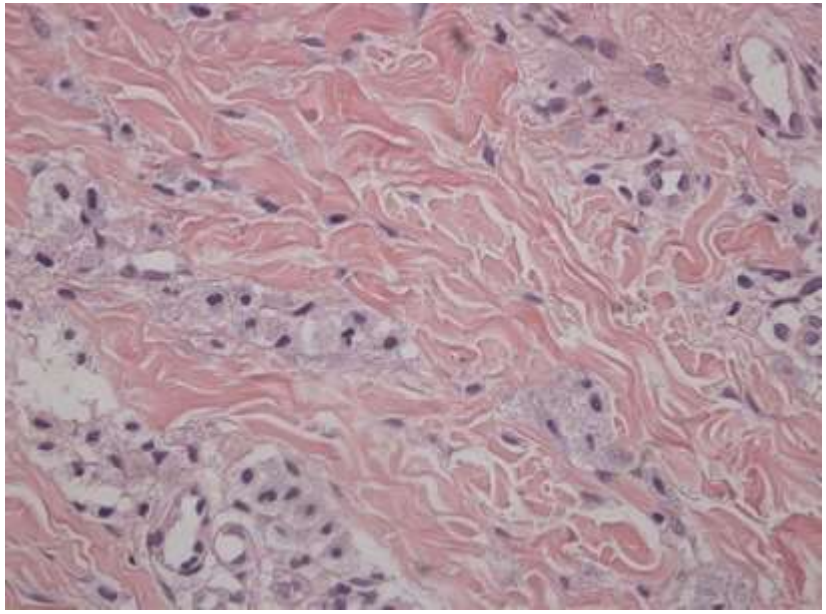


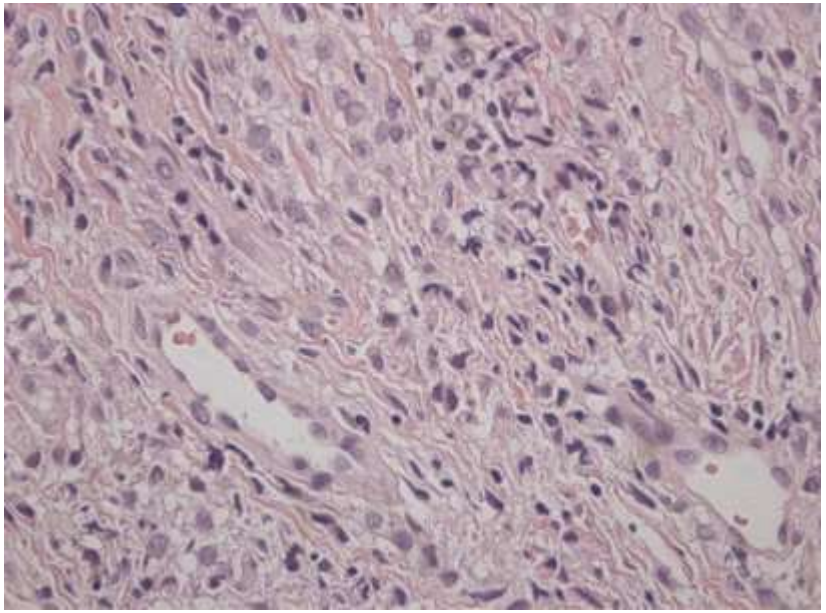
Monster : 2014.1 C	Score
Wat is uw eindconclusie voor Huid III A) Huid zonder afwijkingen B) Huid met mestcelafwijking C) Huid met afwijking, vervolgonderzoek m.b.v. Immunohistochemie geadviseerd D) Niet te beoordelen vanwege kwaliteit coupe/kleuring	2

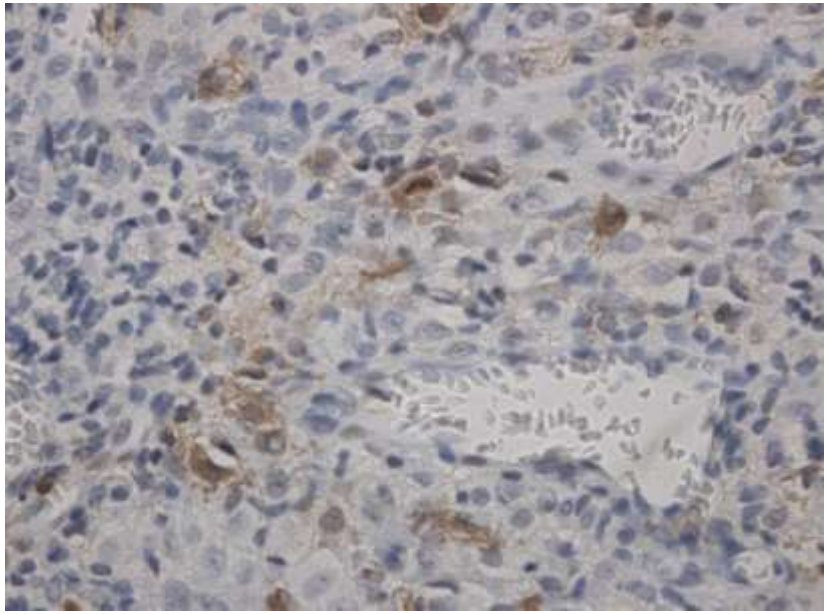


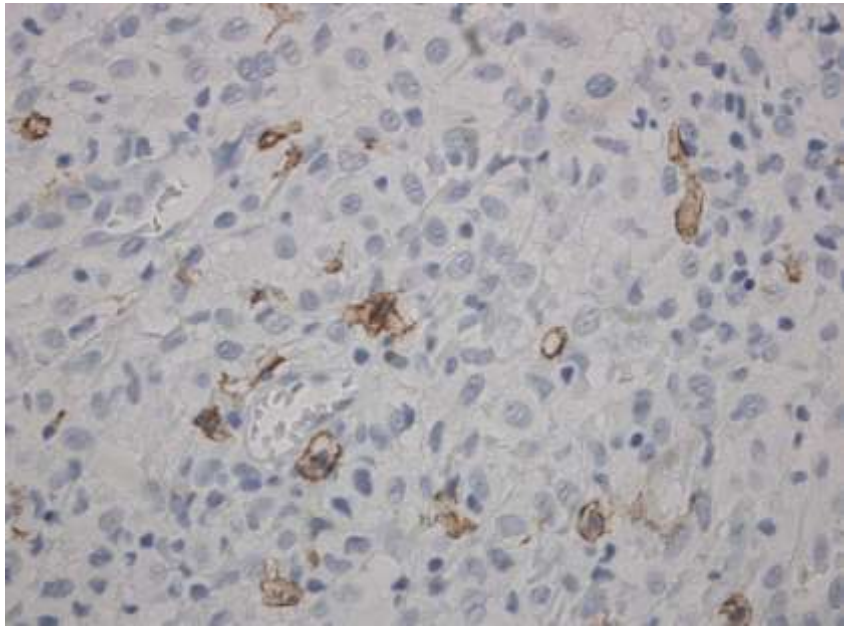


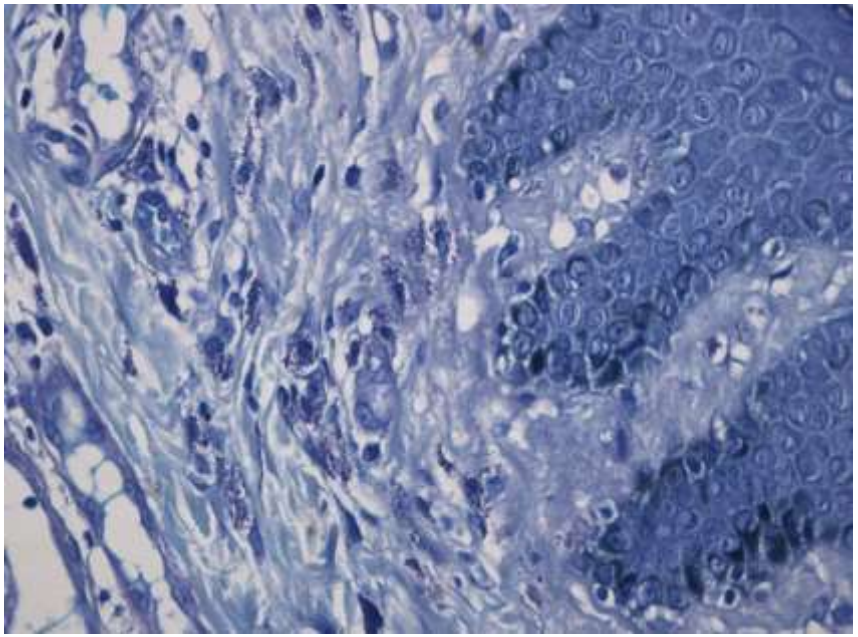


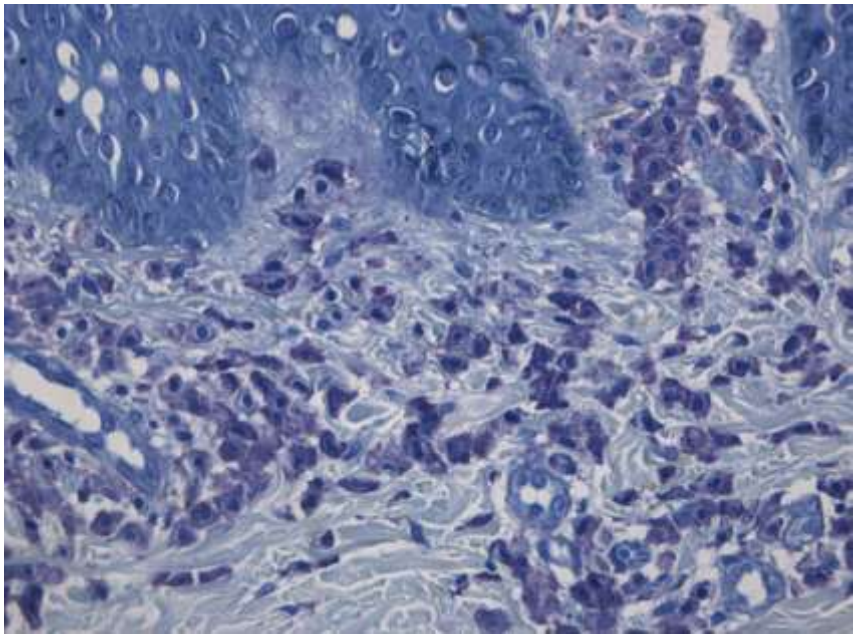


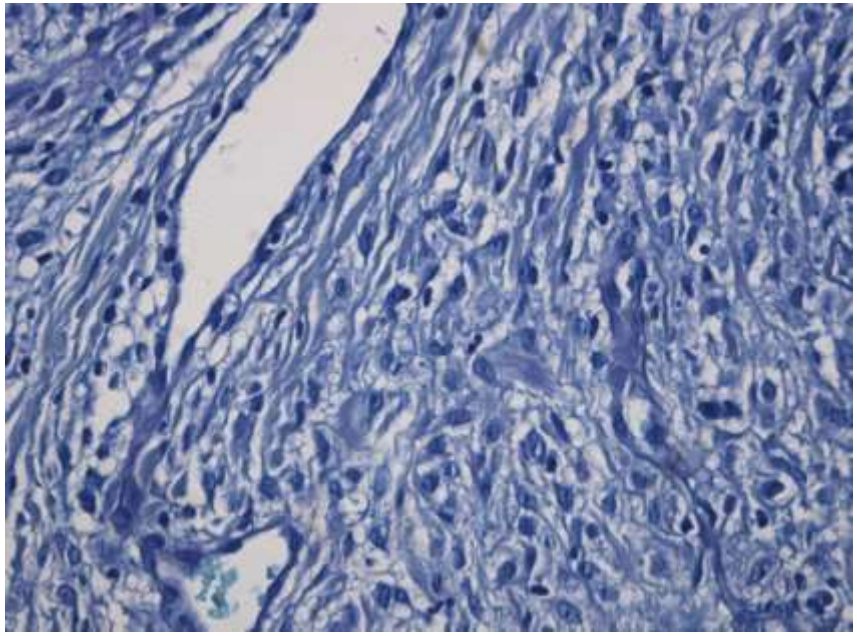


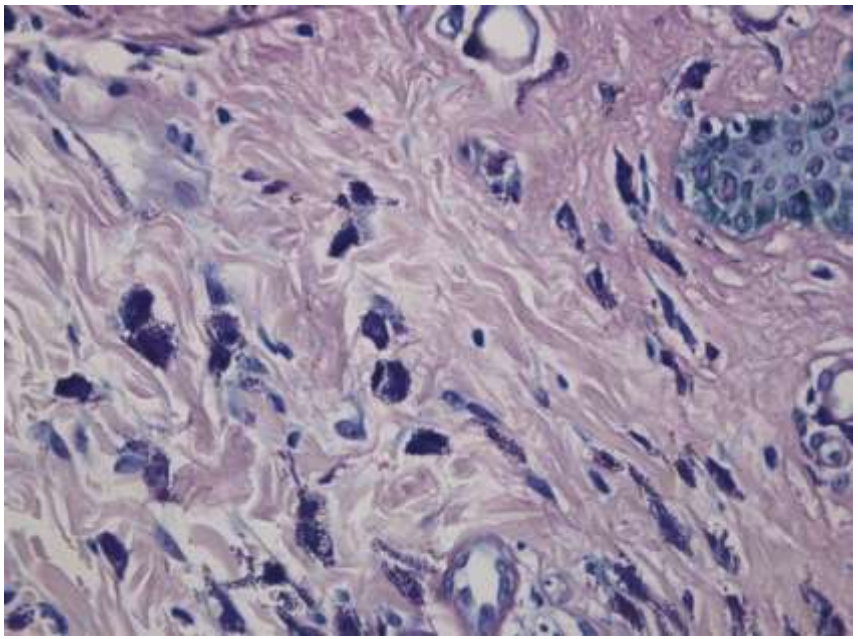


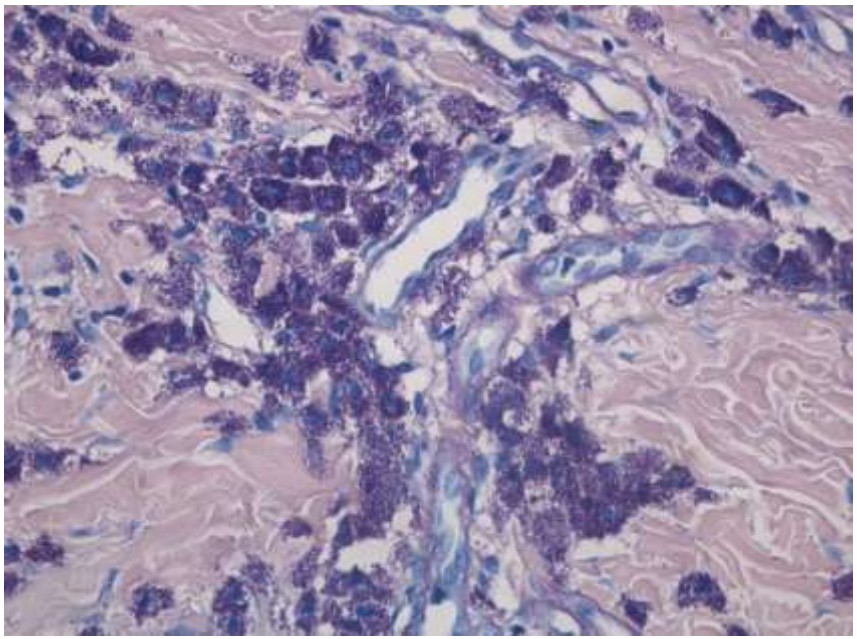


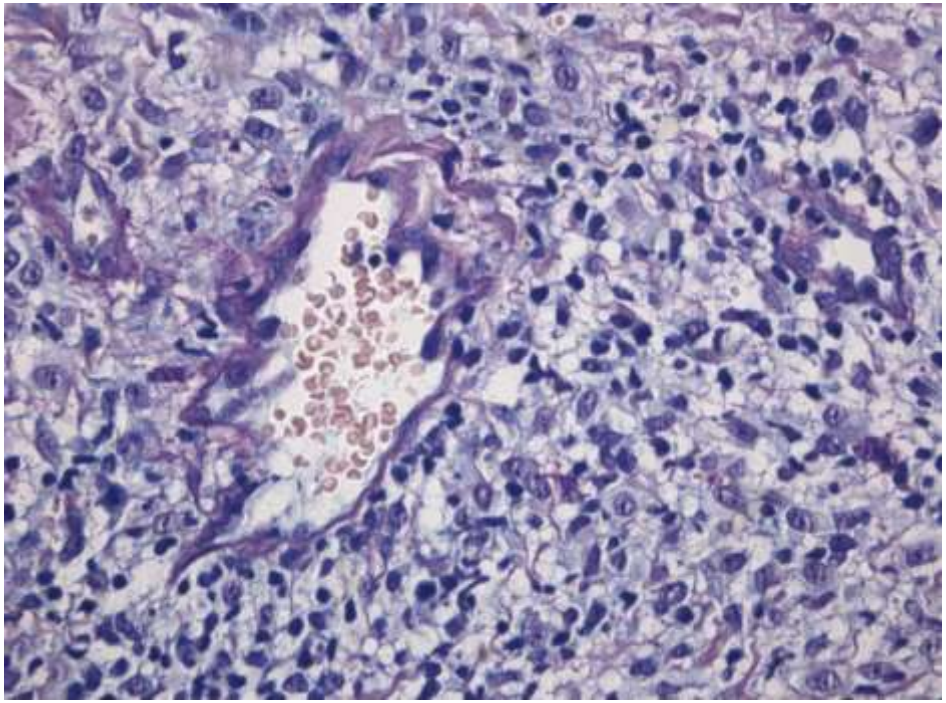


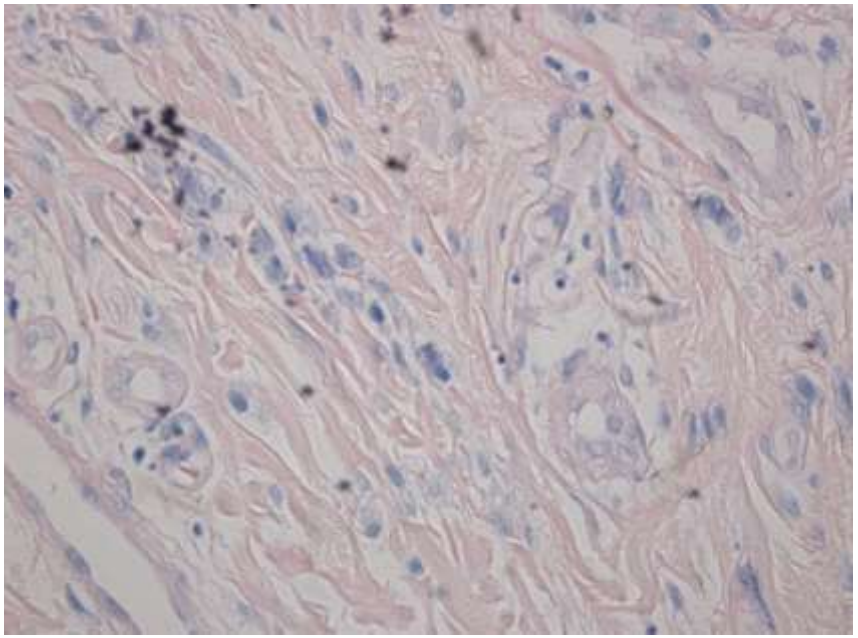


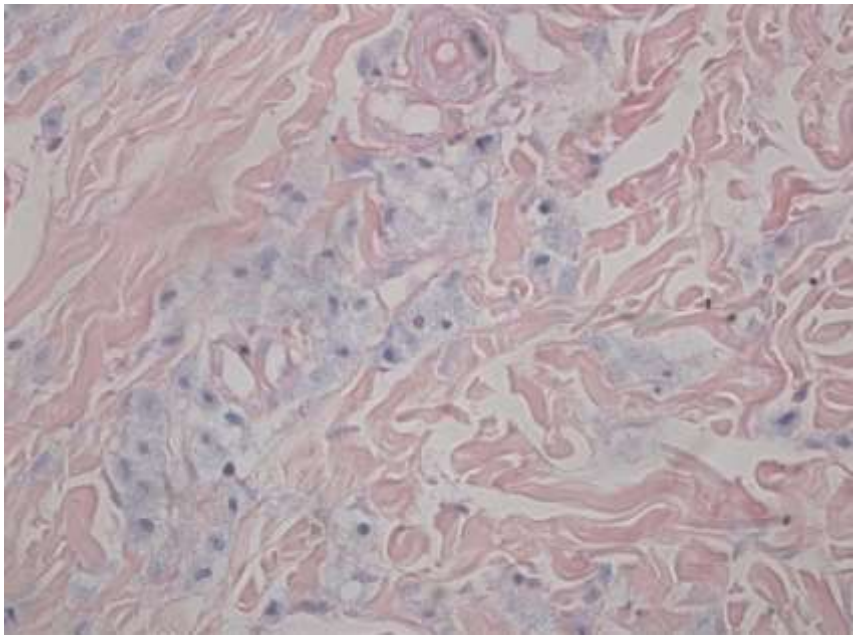


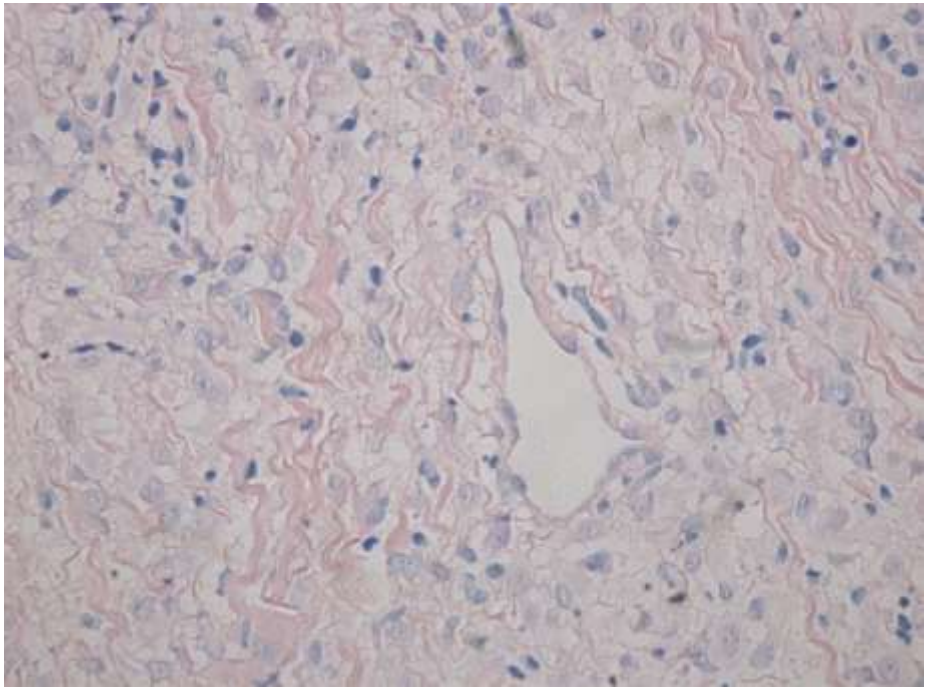


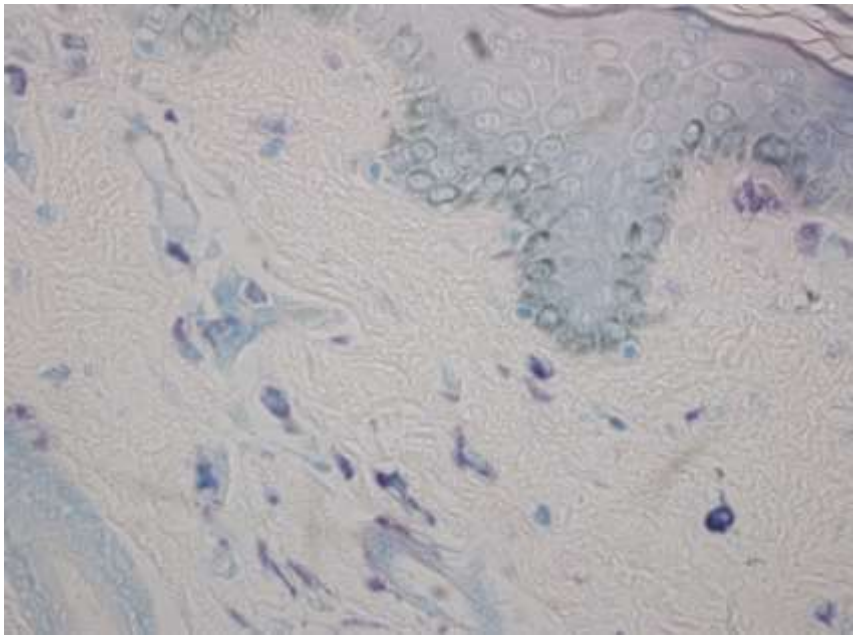


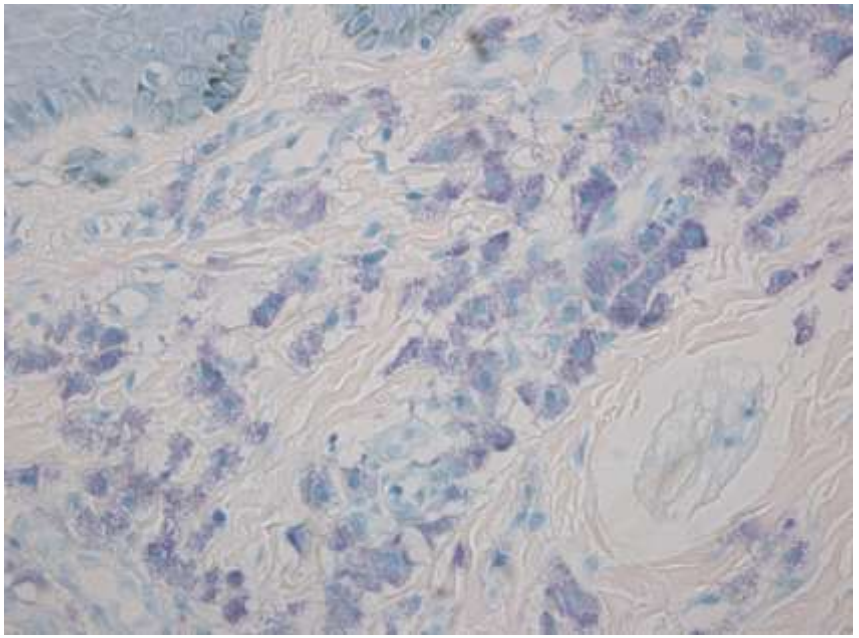


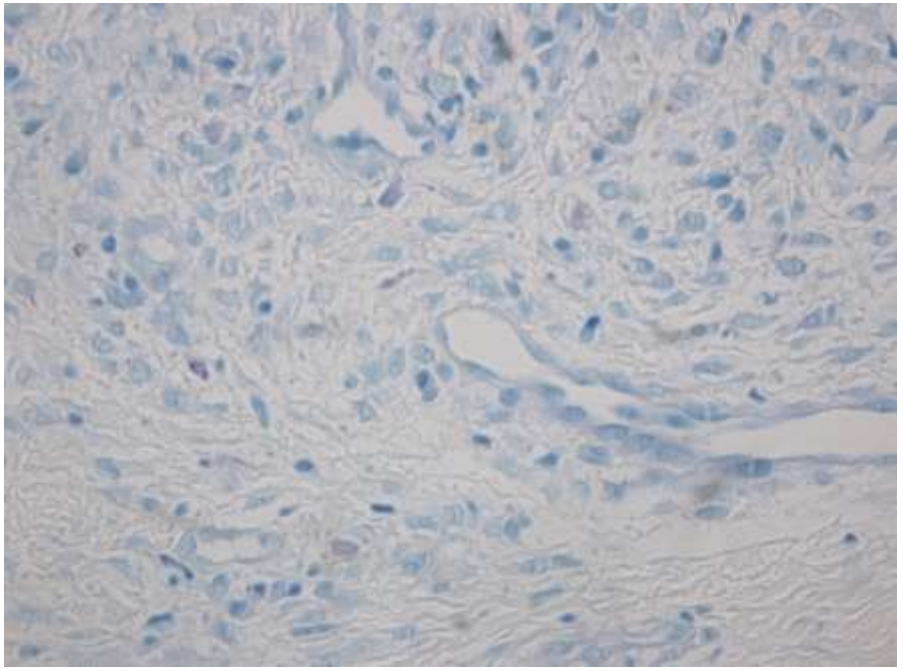


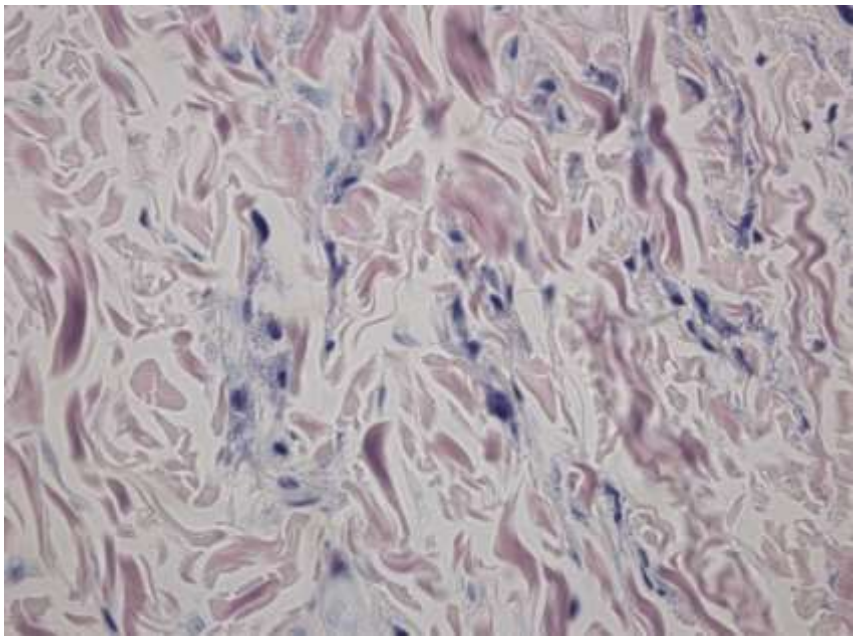


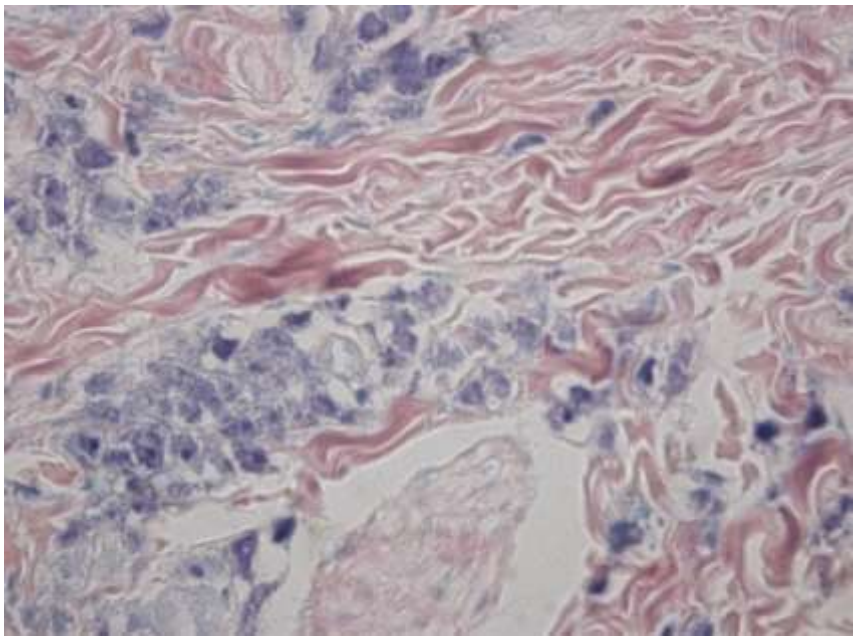


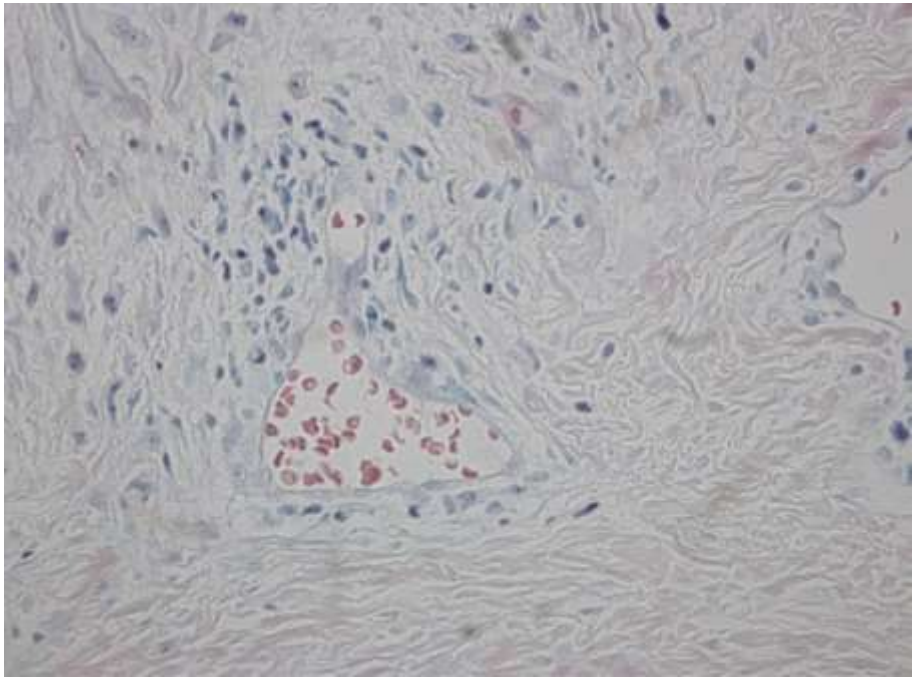


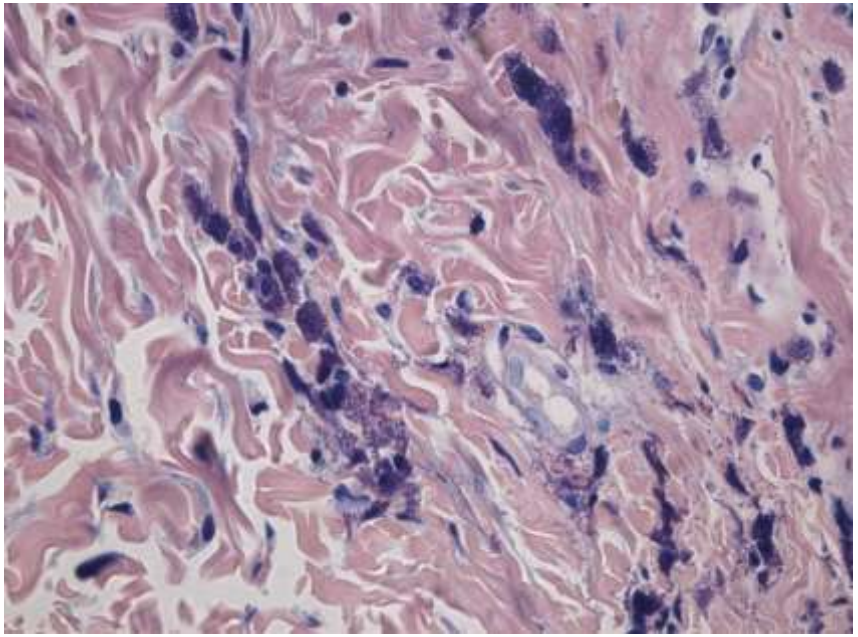


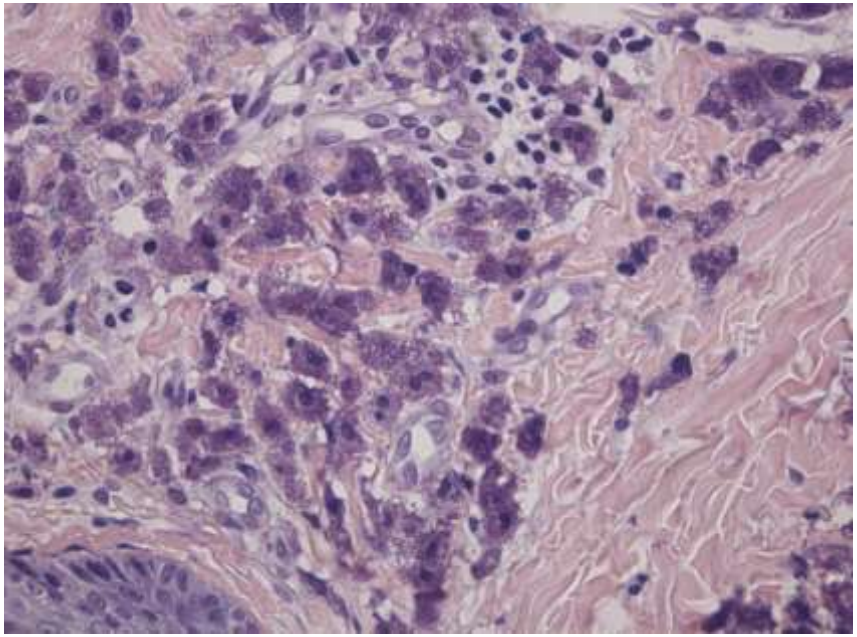


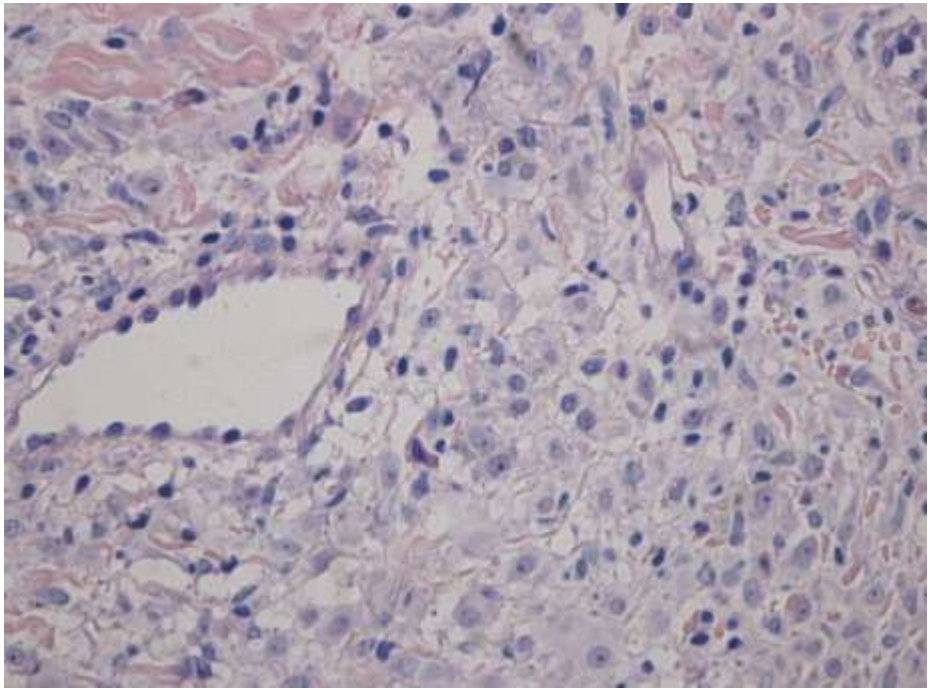


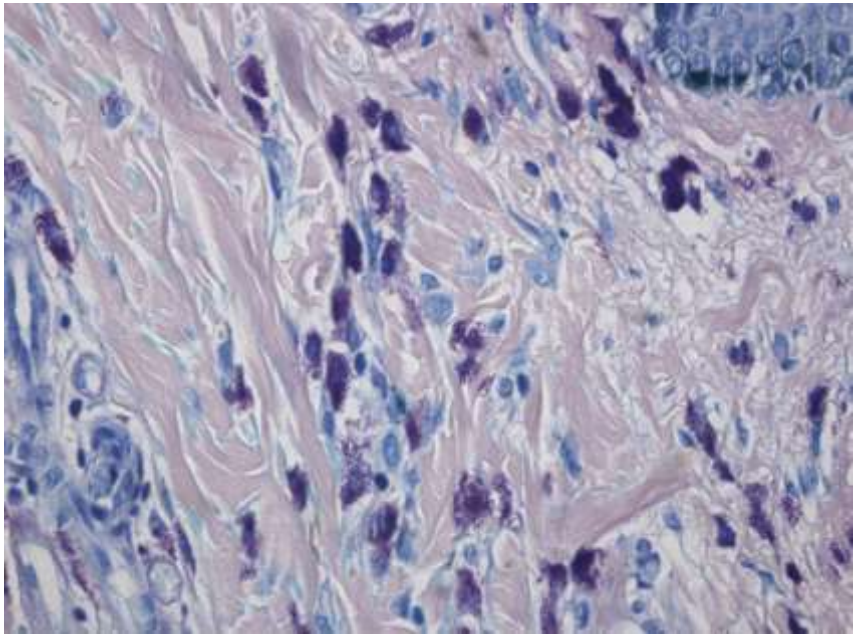


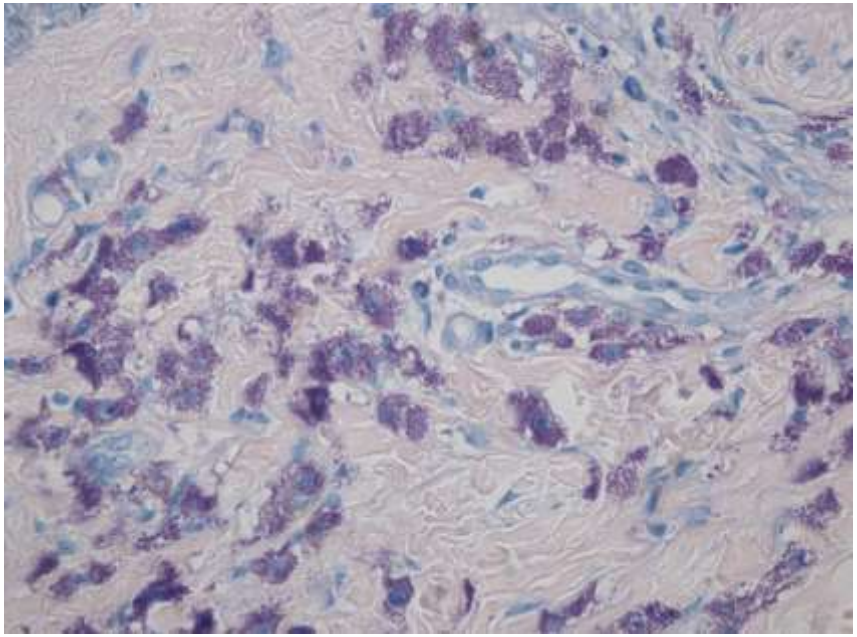


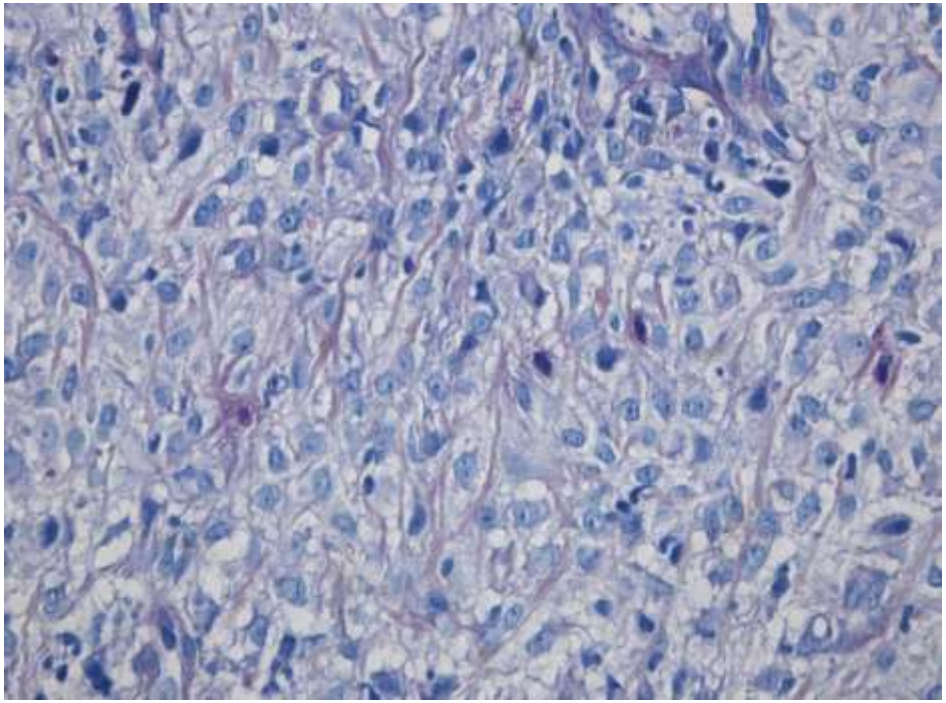












Dankbetuiging

- Fotografie: Fred Vlugt
- Secretariële ondersteuning:
  Miriam Hoenderdos