

AF 45/05 SERCE
083387 80.0KV X3000

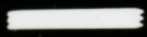
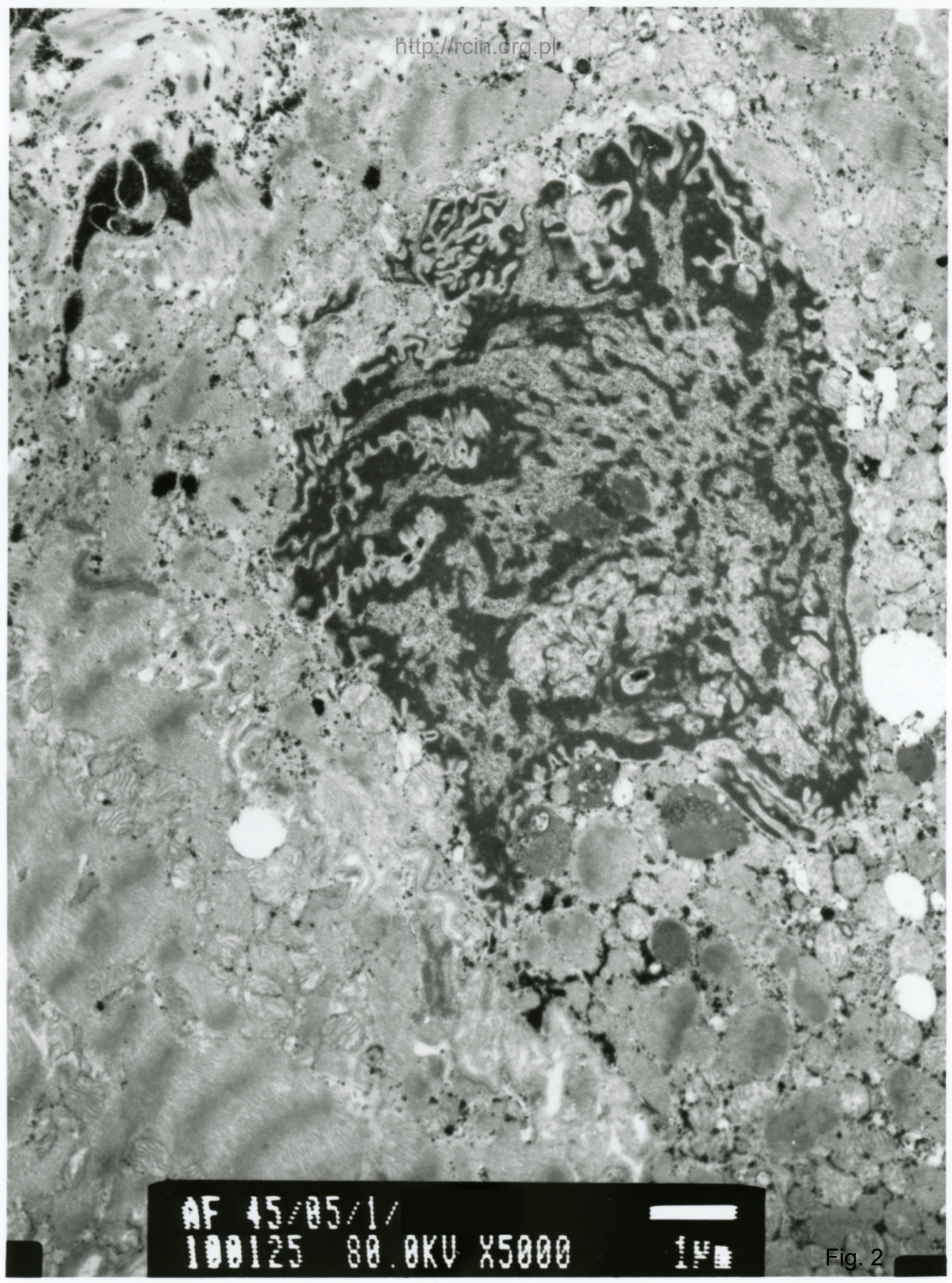
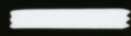


Fig. 1

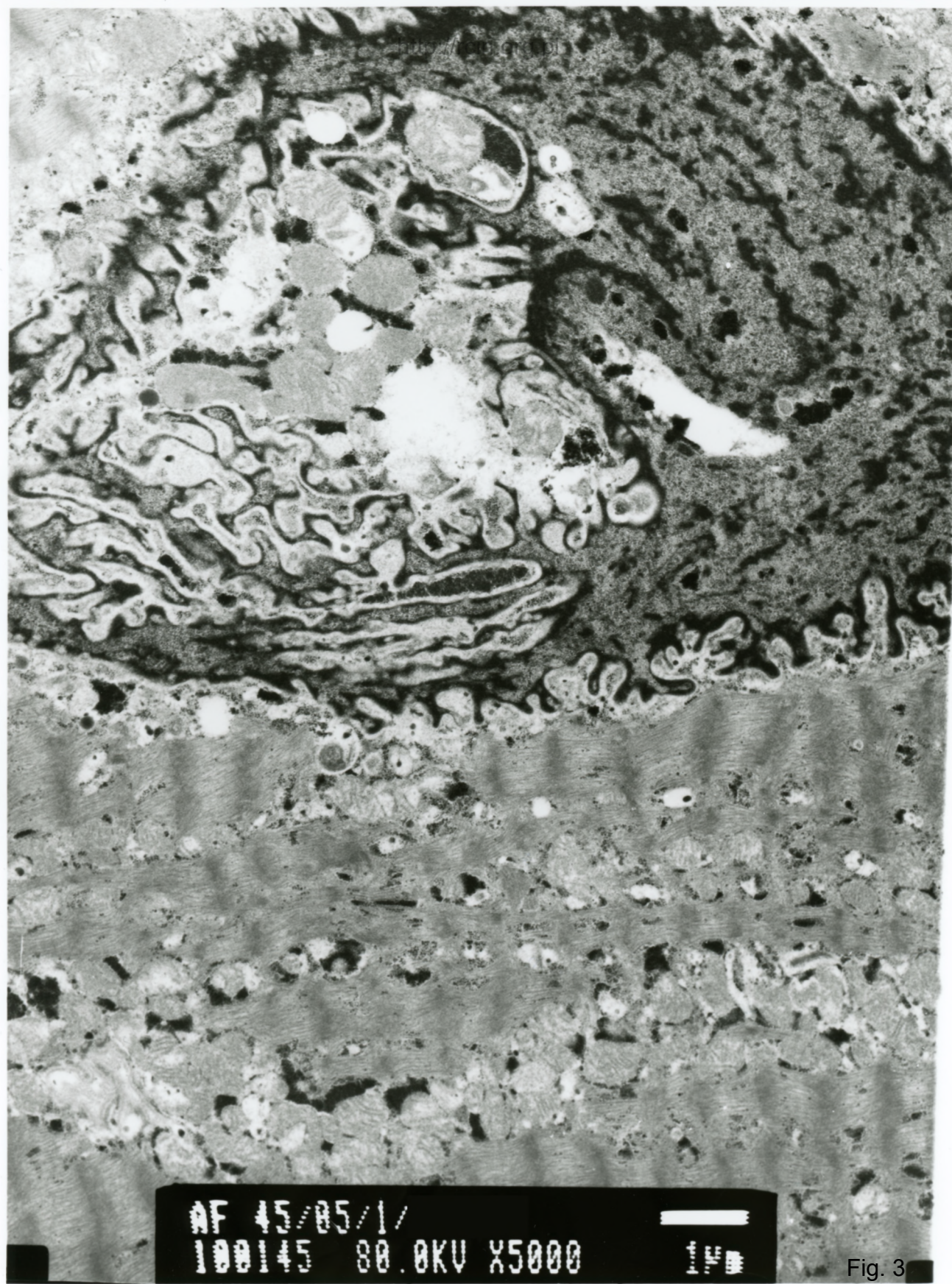


AF 45/05/1/
100125 80.0KV X5000



1 μm

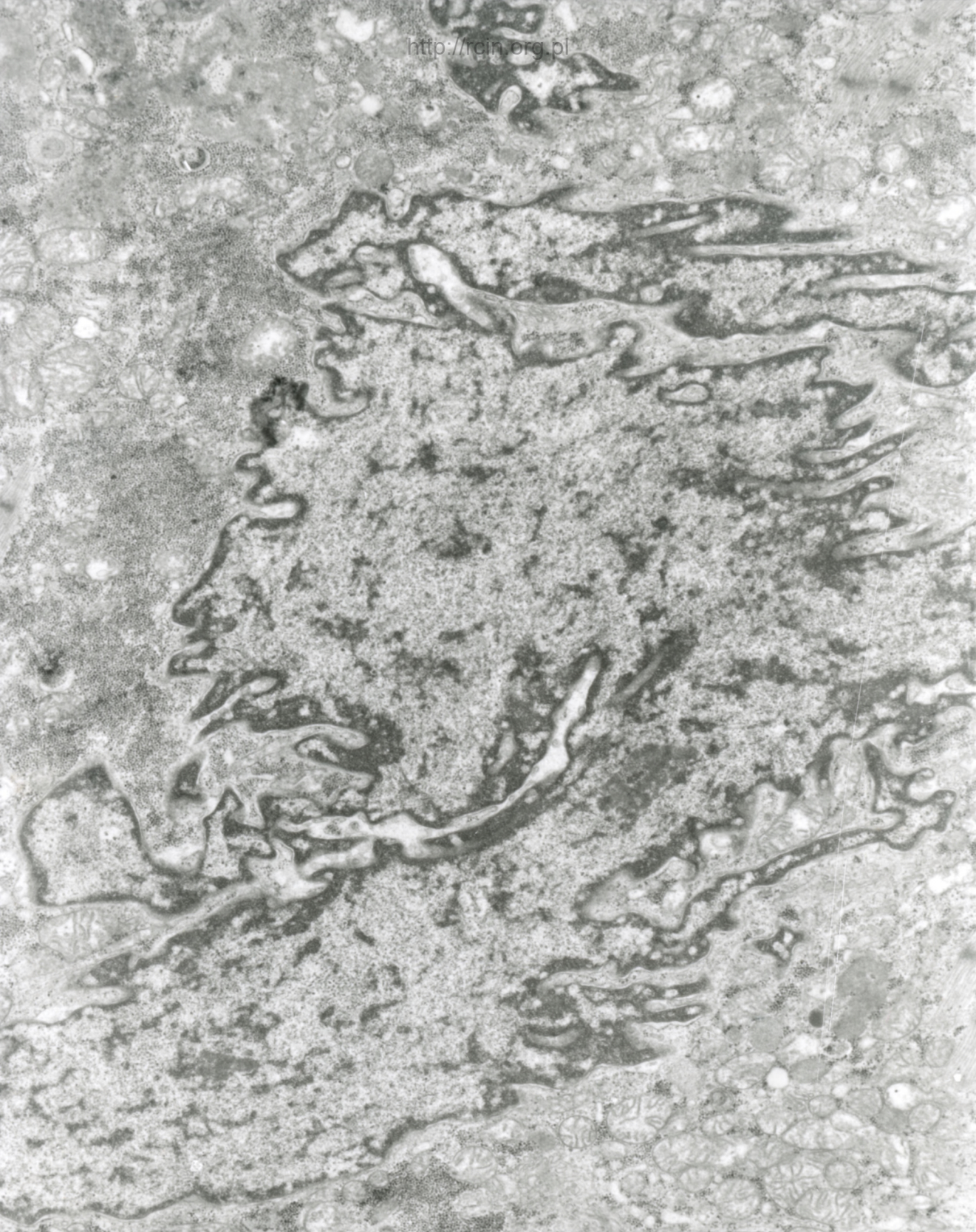
Fig. 2



AF 45/05/1/
100145 80.0KV X5000

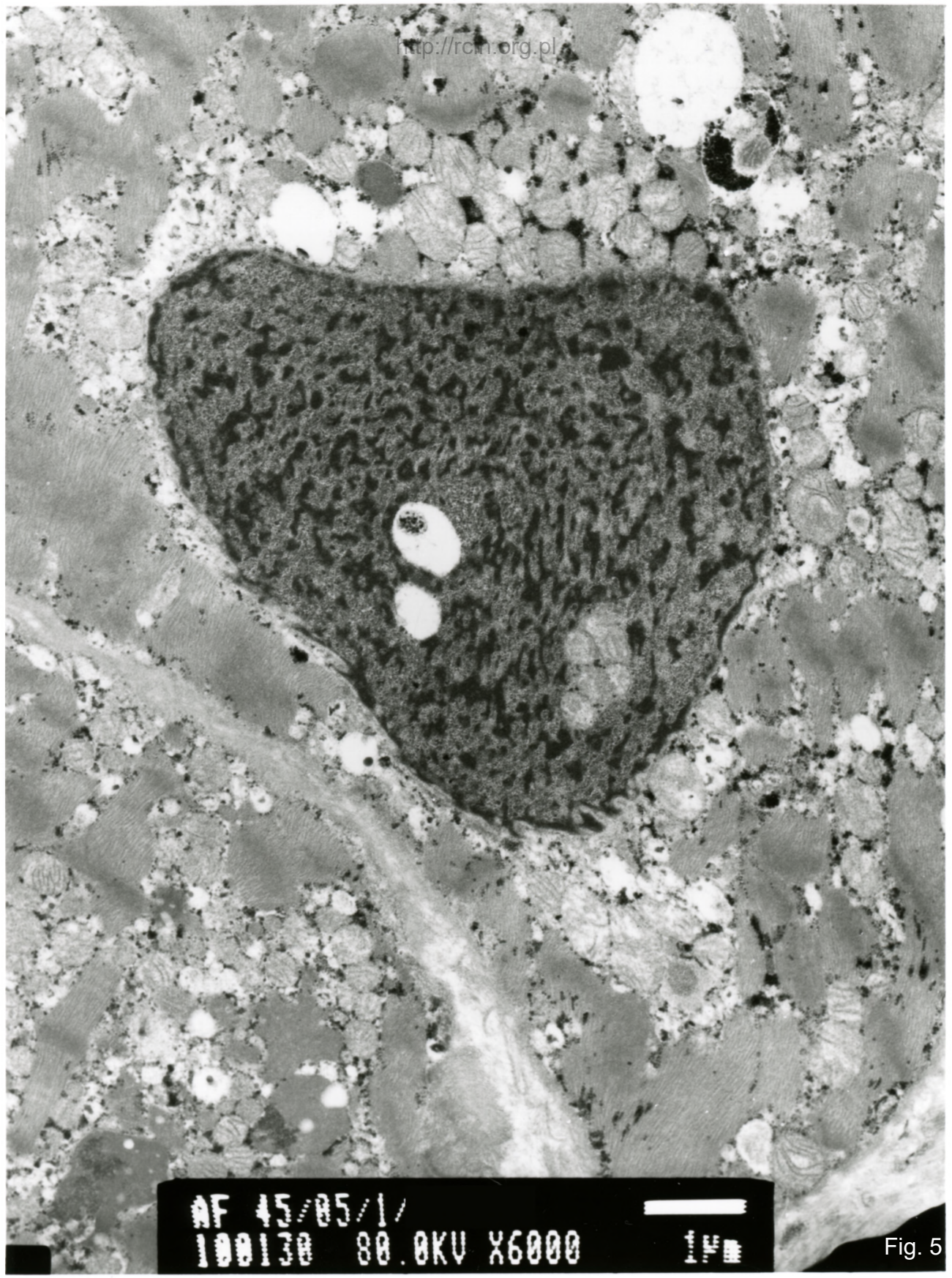


Fig. 3



AF 45/05/SERCR
050510 80.0KV X5000

Fig. 4

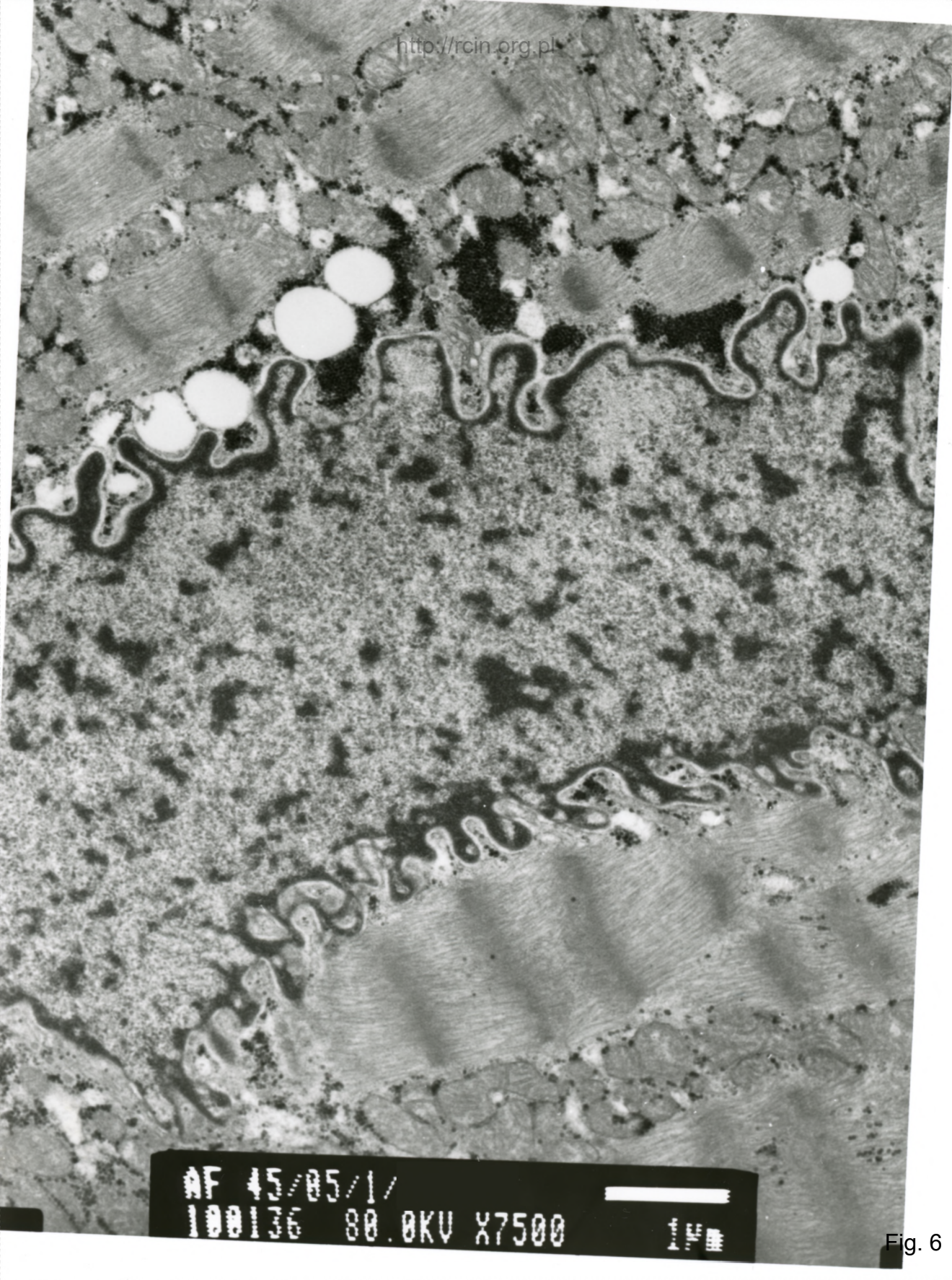


AF 45/05/1/
100130 80.0KV X6000



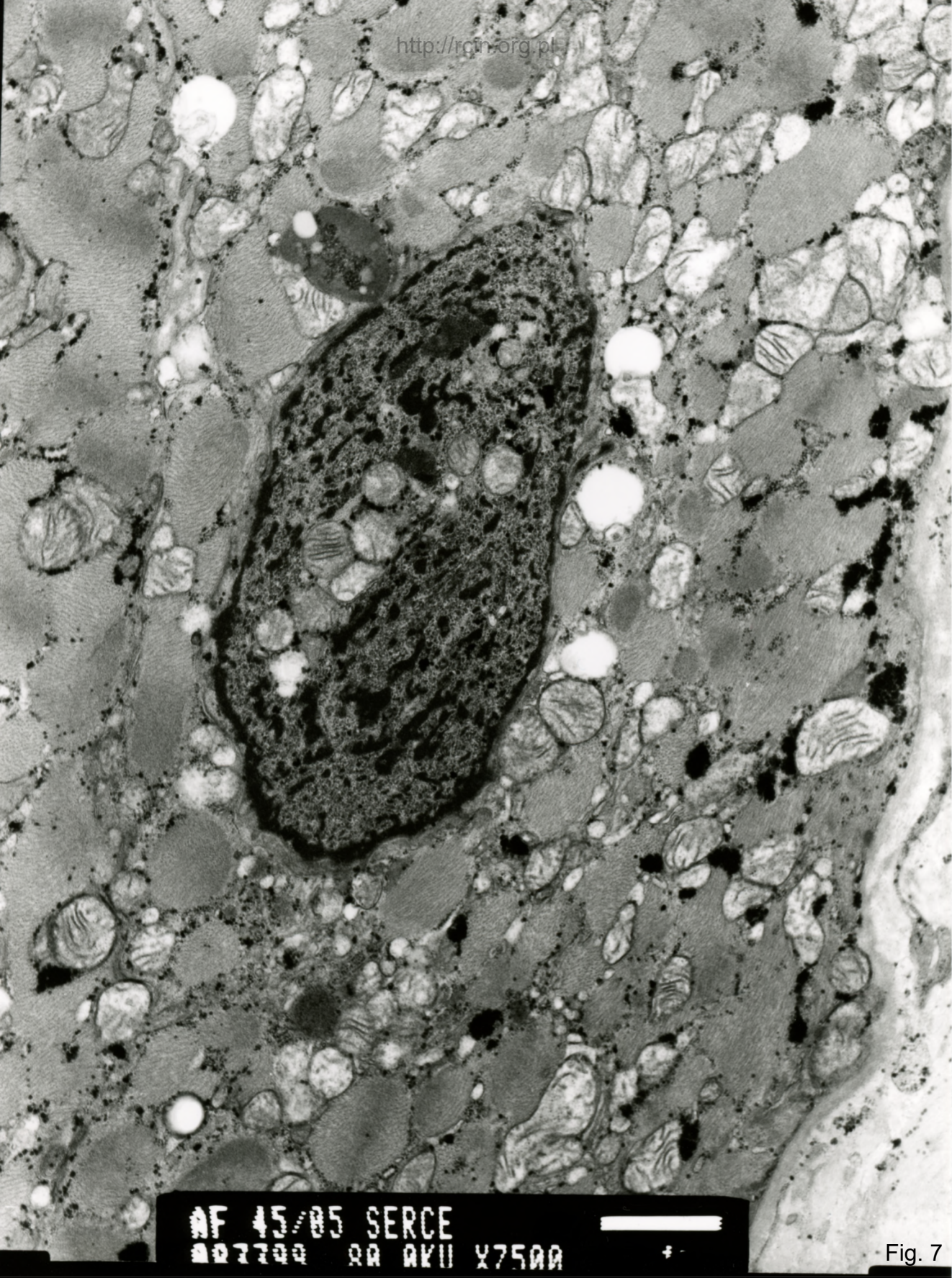
1 μm

Fig. 5



AF 45/05/1/
100136 80.0KV X7500

Fig. 6



MF 45/05 SERCE
007700 RR RKII X7500

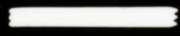
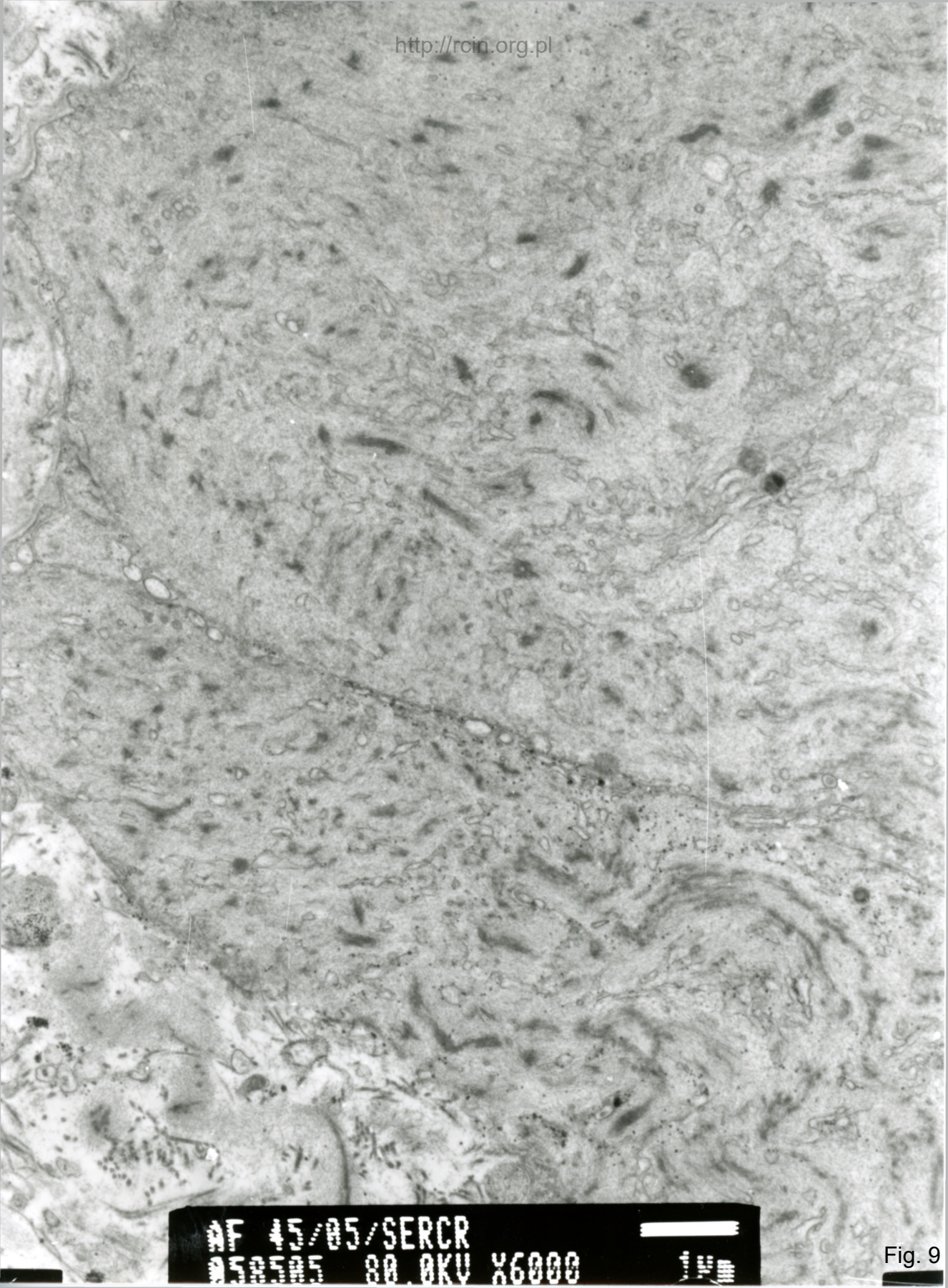


Fig. 7



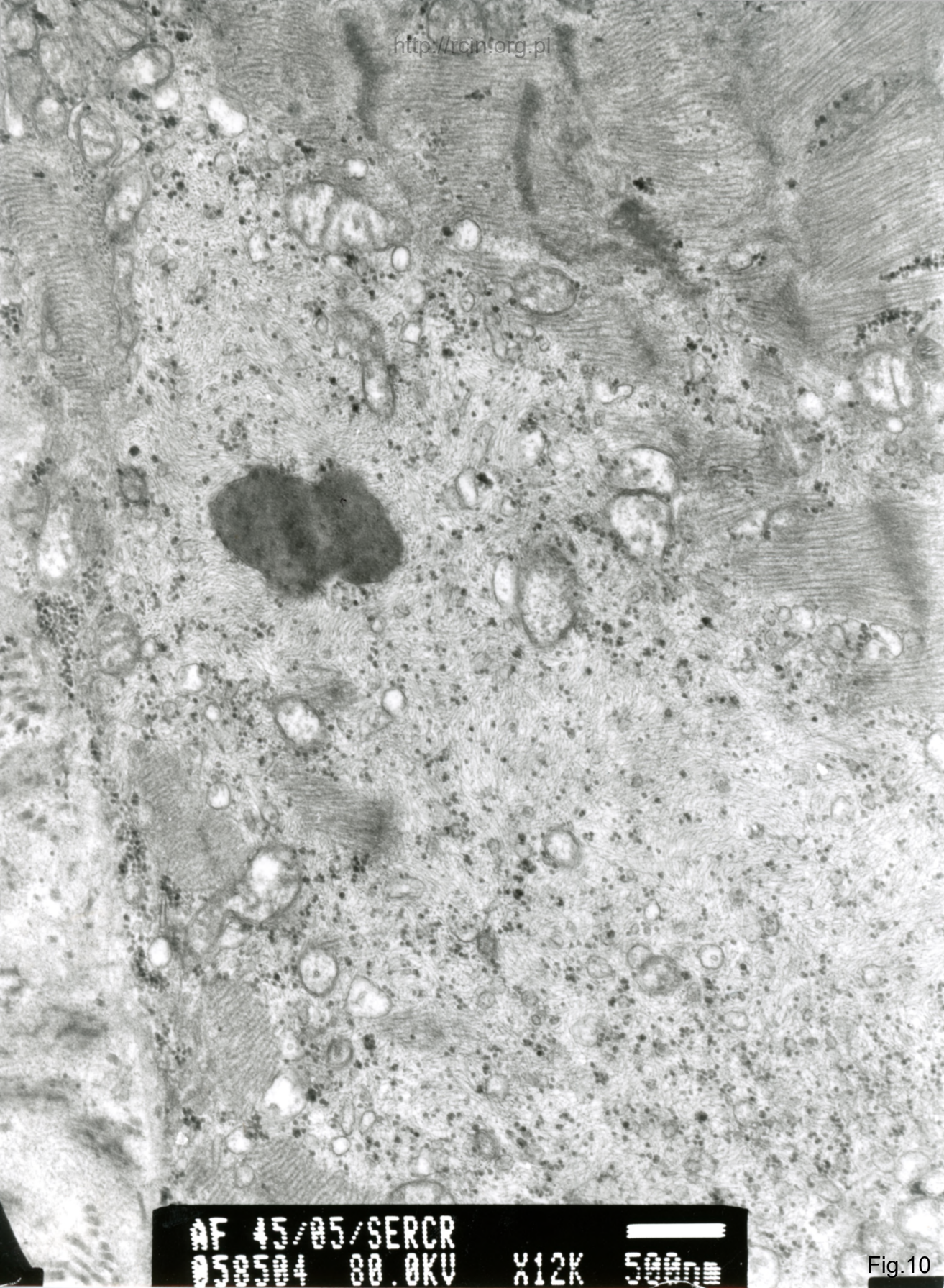
AF 45/05 SERCE
007100 8A AKU X15K 500

Fig. 8



AF 15/05/SERCR
050505 80.0KV X6000

Fig. 9



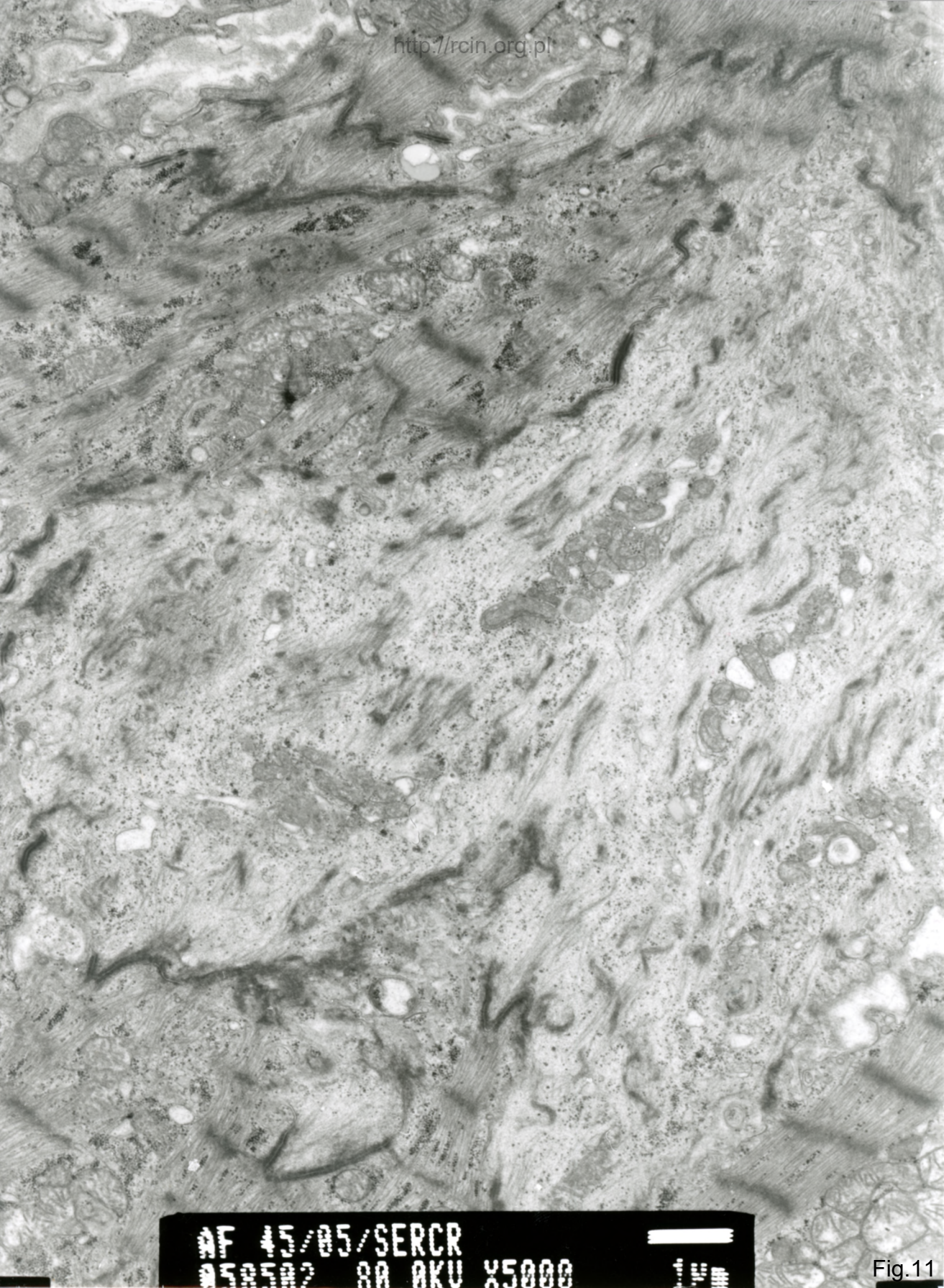
AF 45/05/SERCR

050504 80.0KV

X12K

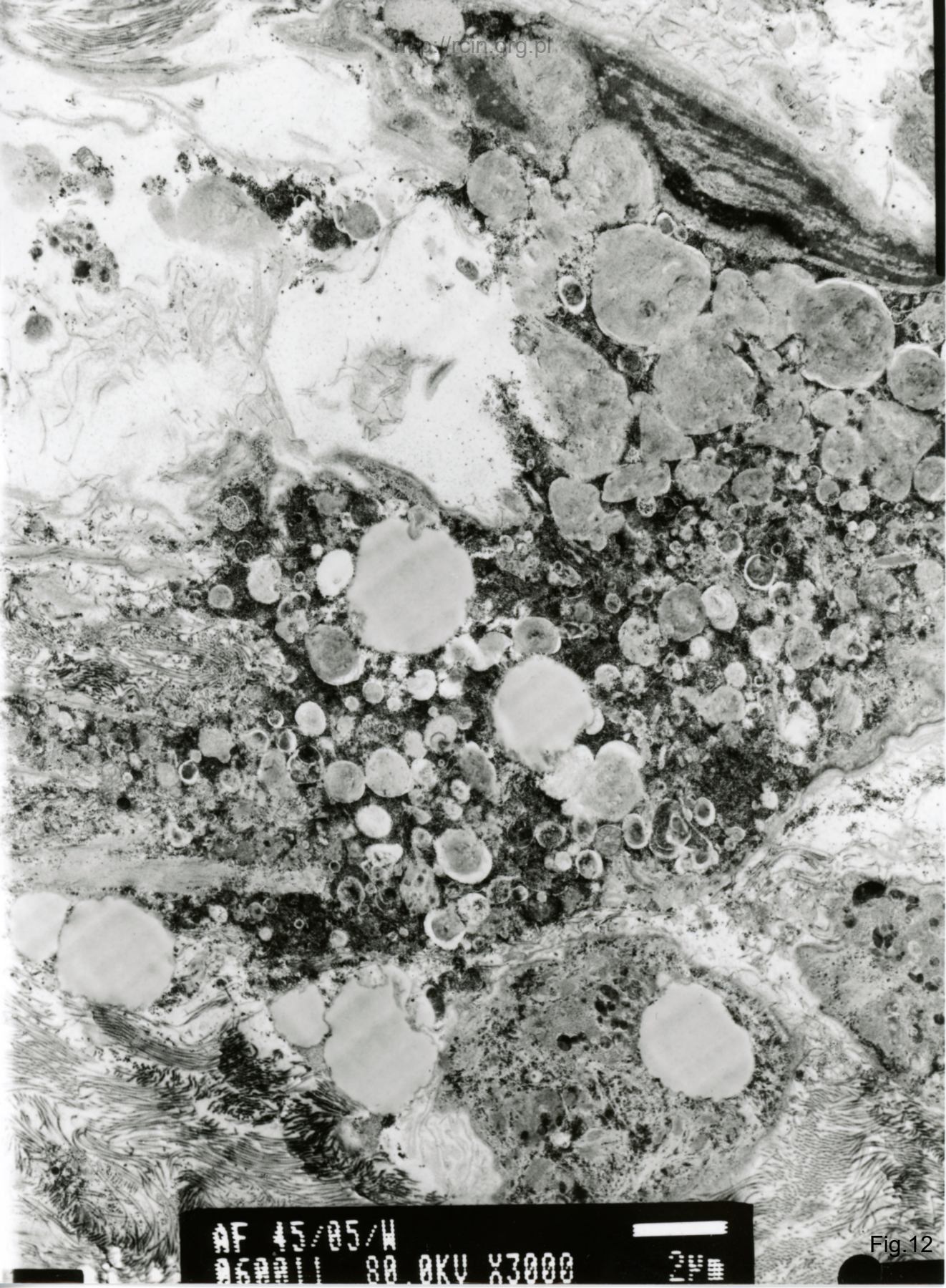
500nm

Fig.10



AF 45/05/SERCR
050502 80 AKU X5000

Fig.11



AF 45/05/W
060111 80.0KV X3000 24

Fig.12

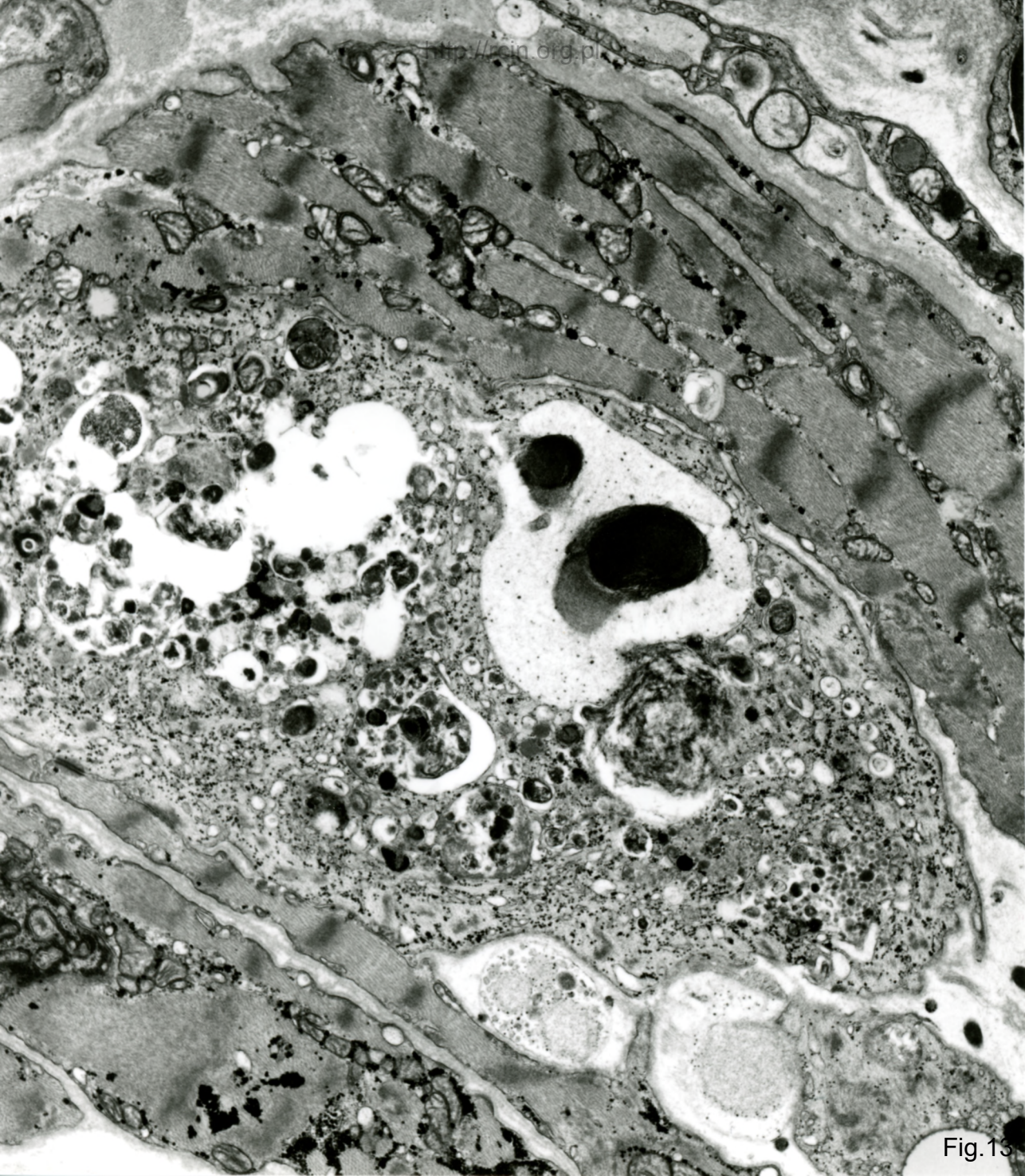
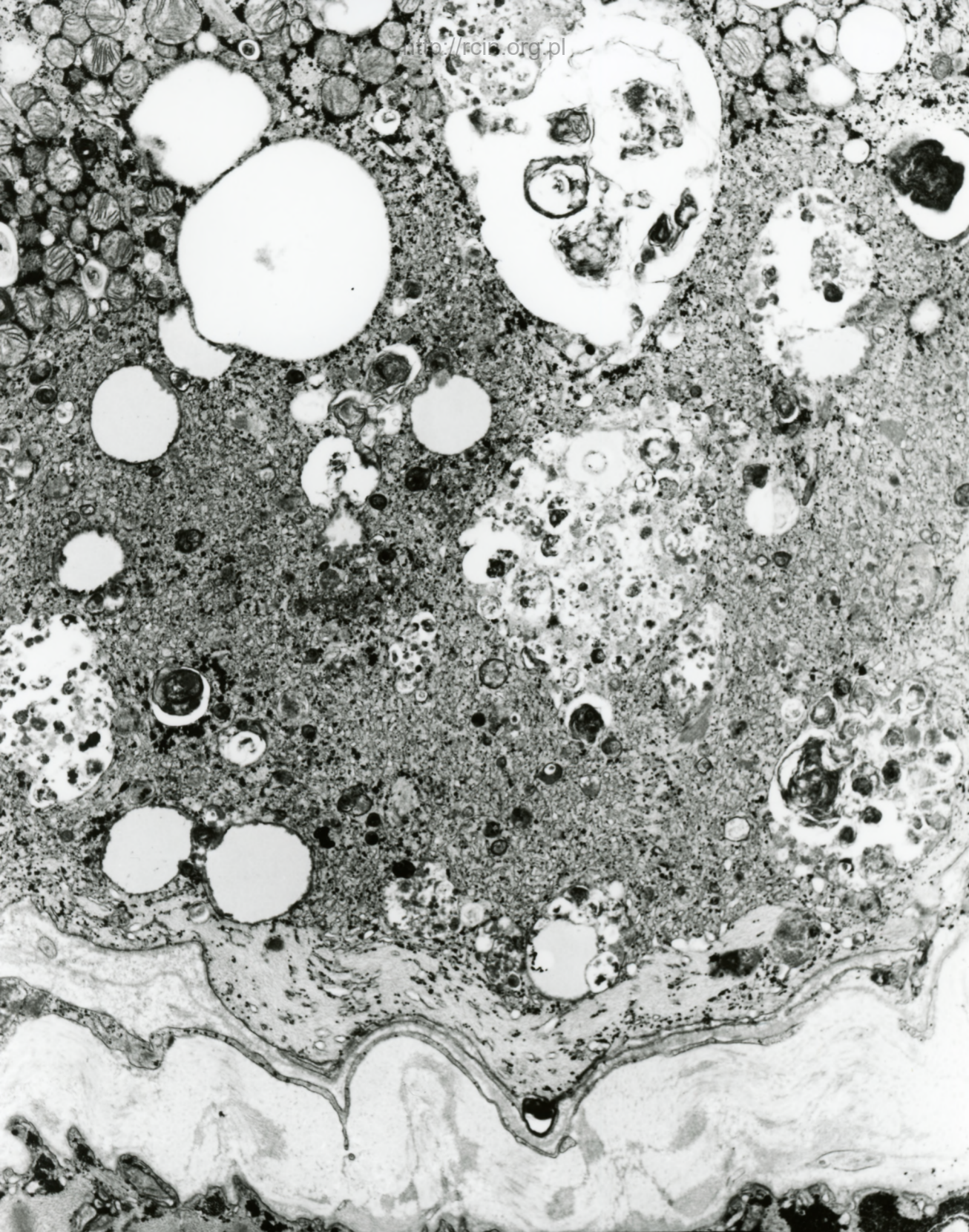
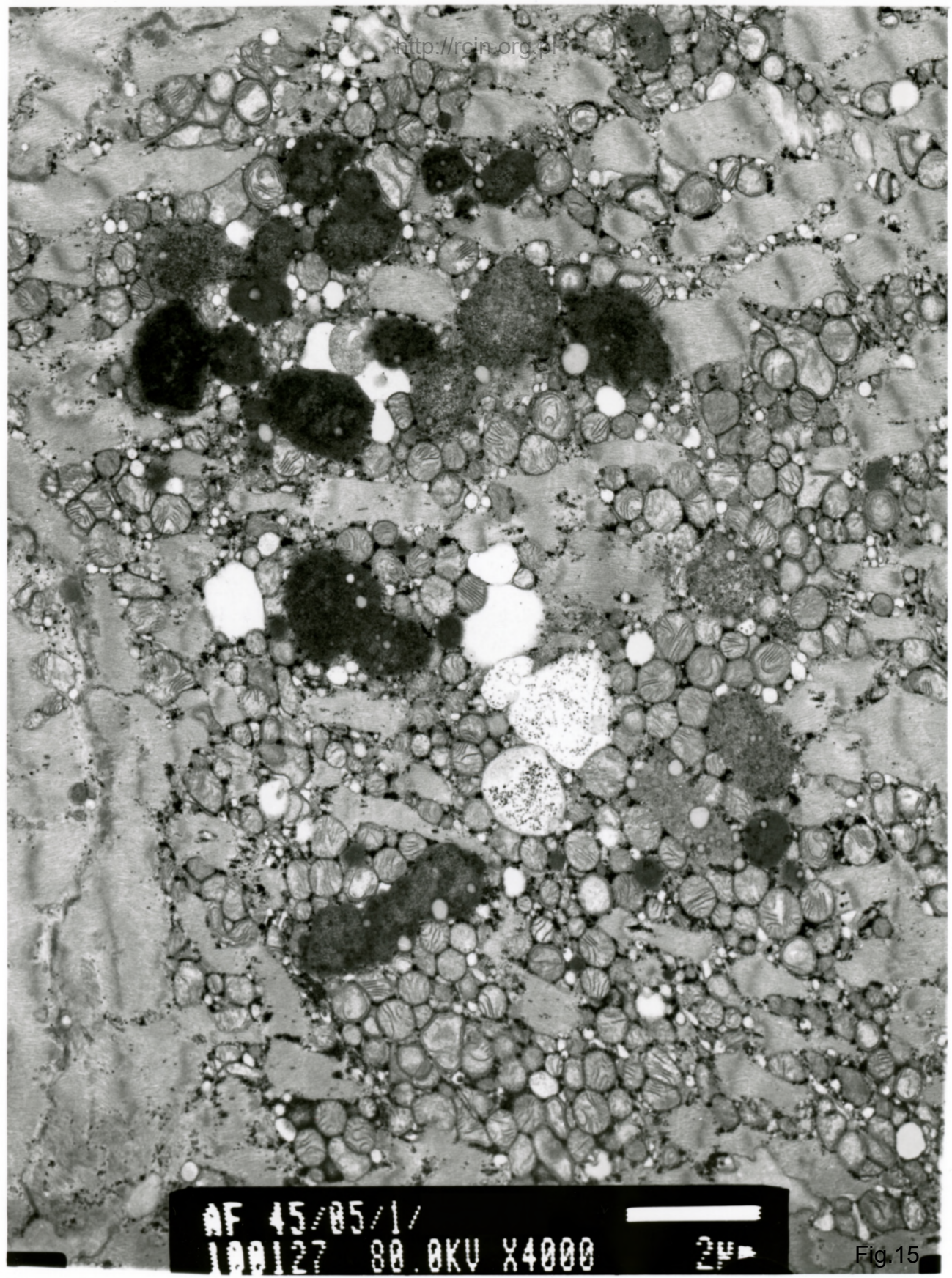


Fig. 13



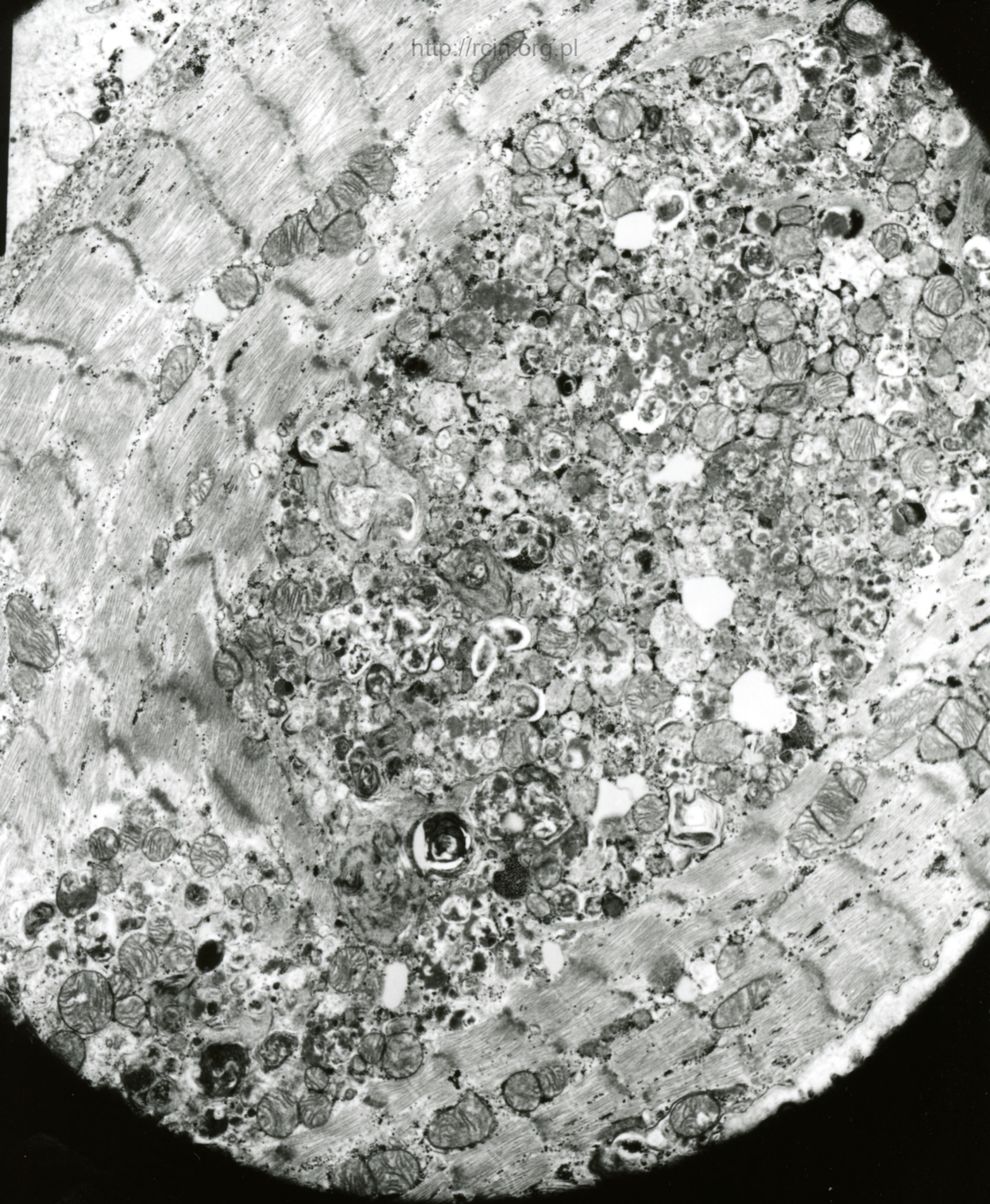
AF 45/05
003369 80 0KU X4000

Fig. 14



AF 45/05/1/
100127 80.0KV X4000 2μ

Fig. 15



NF 45/05

003367 80 0KV X4000

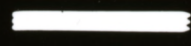
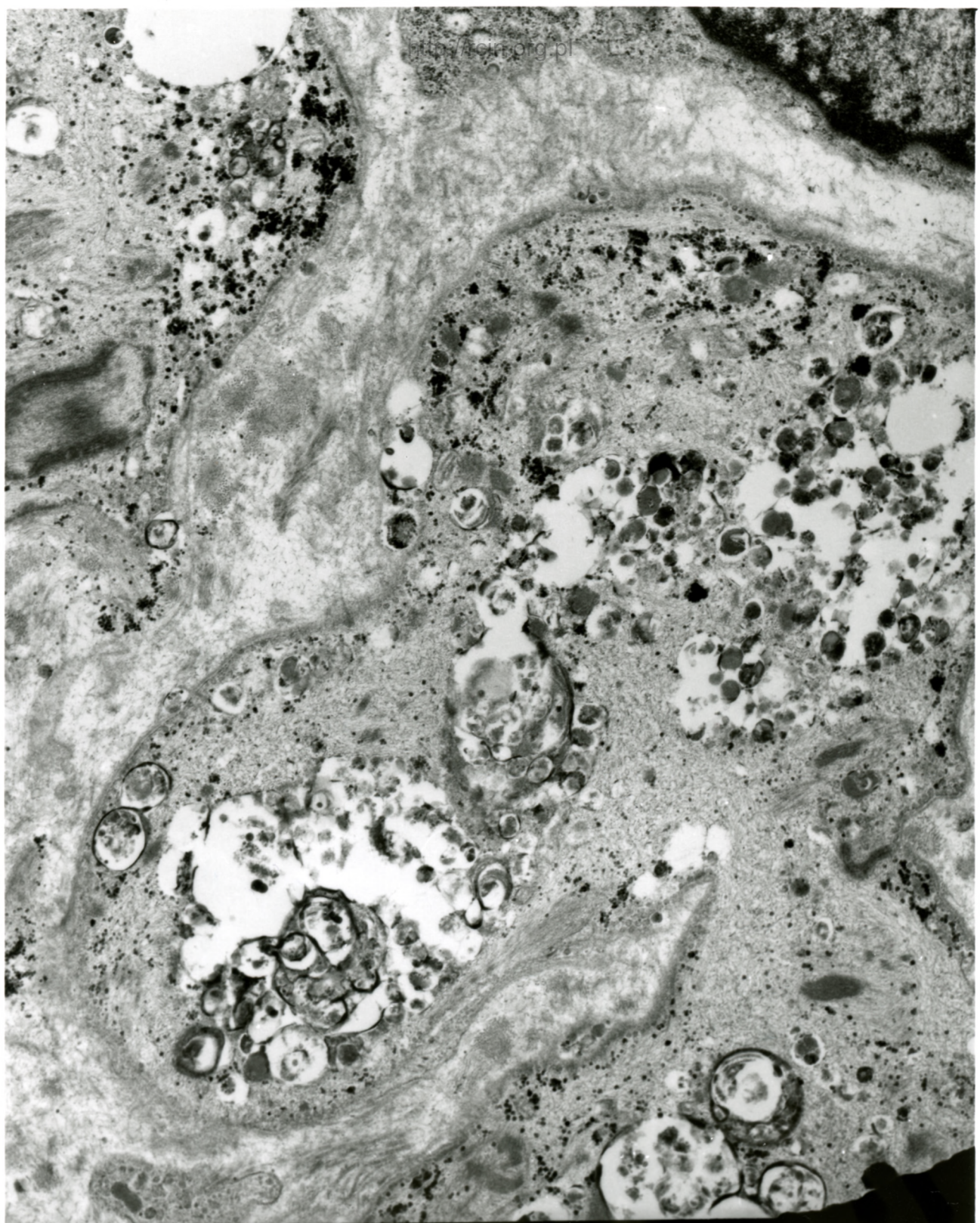


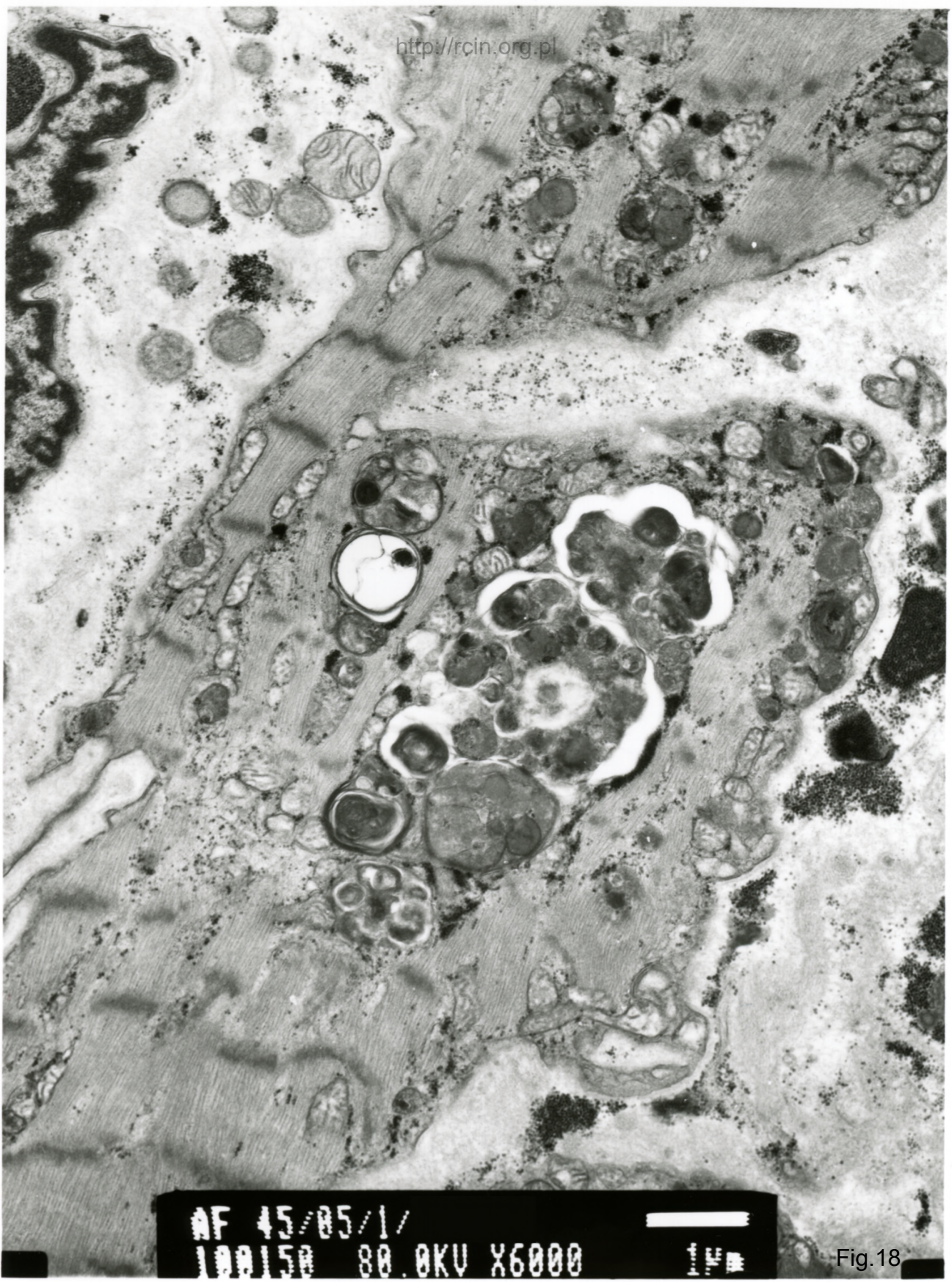
Fig.16



AF 45/05/1/
100133 80.0KV X6000



Fig.17



AF 45/05/1/
100150 80.0KV X6000

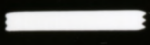
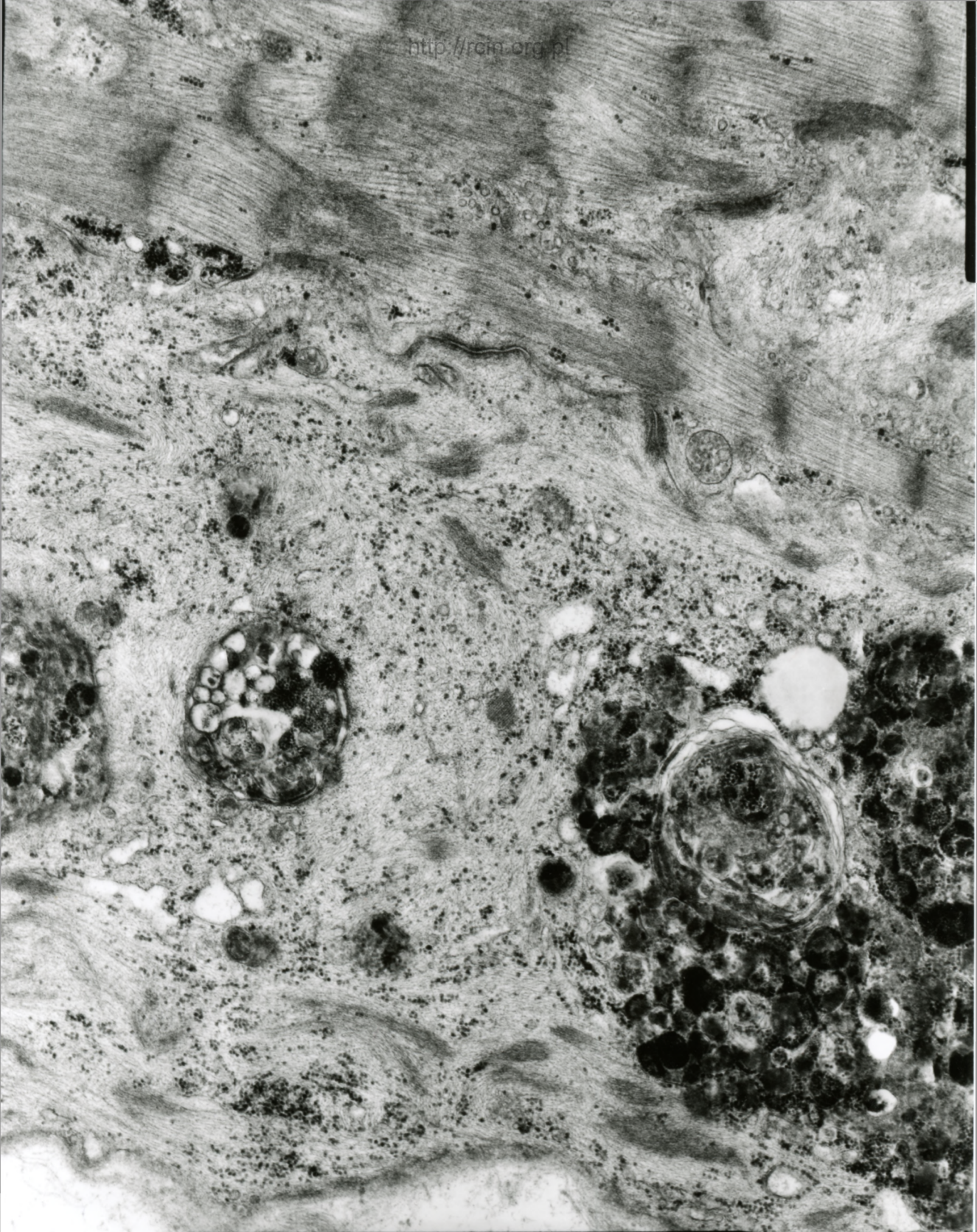
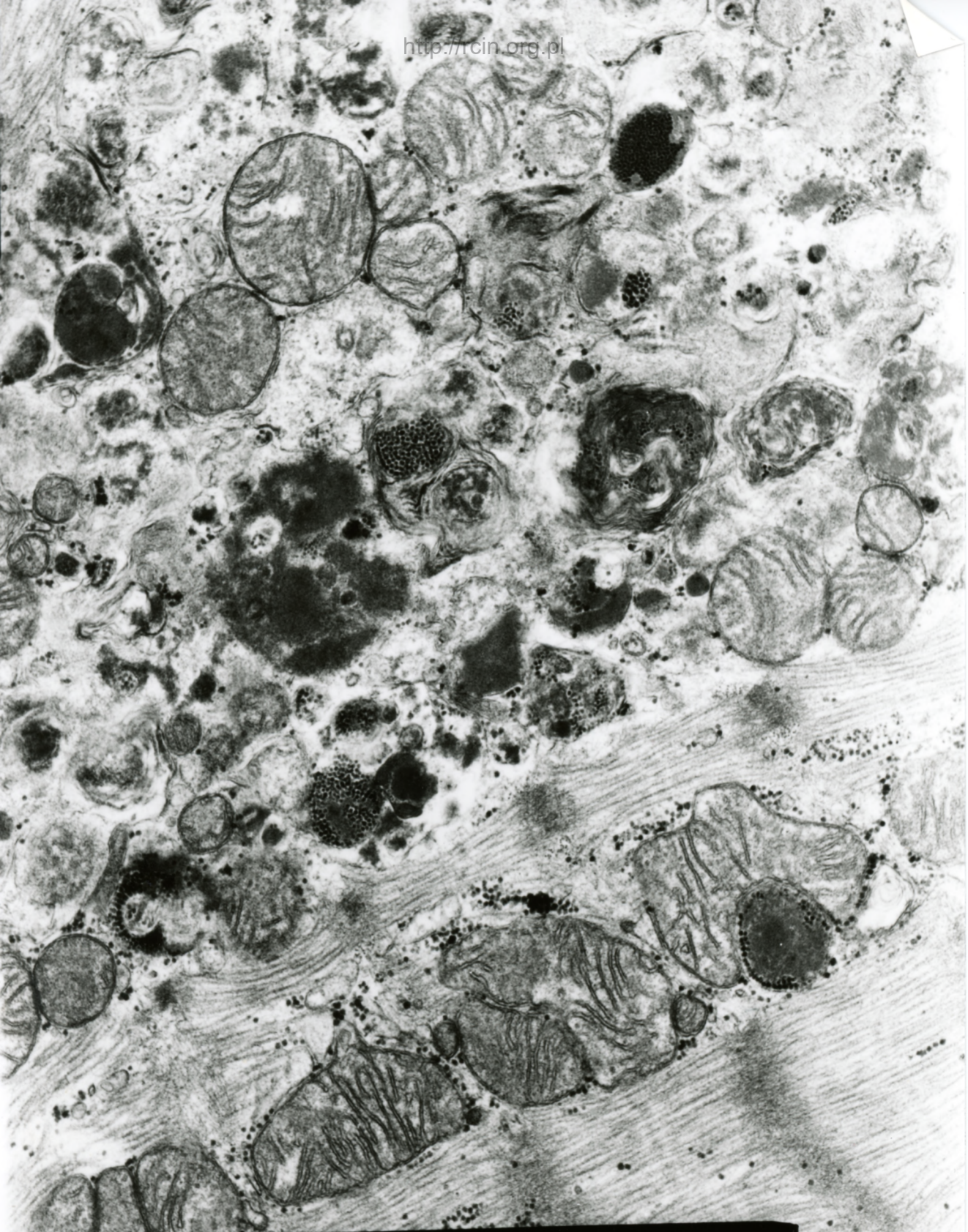


Fig.18



AF 45/05/W
060015 80.0KV X10K 500nm

Fig.19



AF 45/85

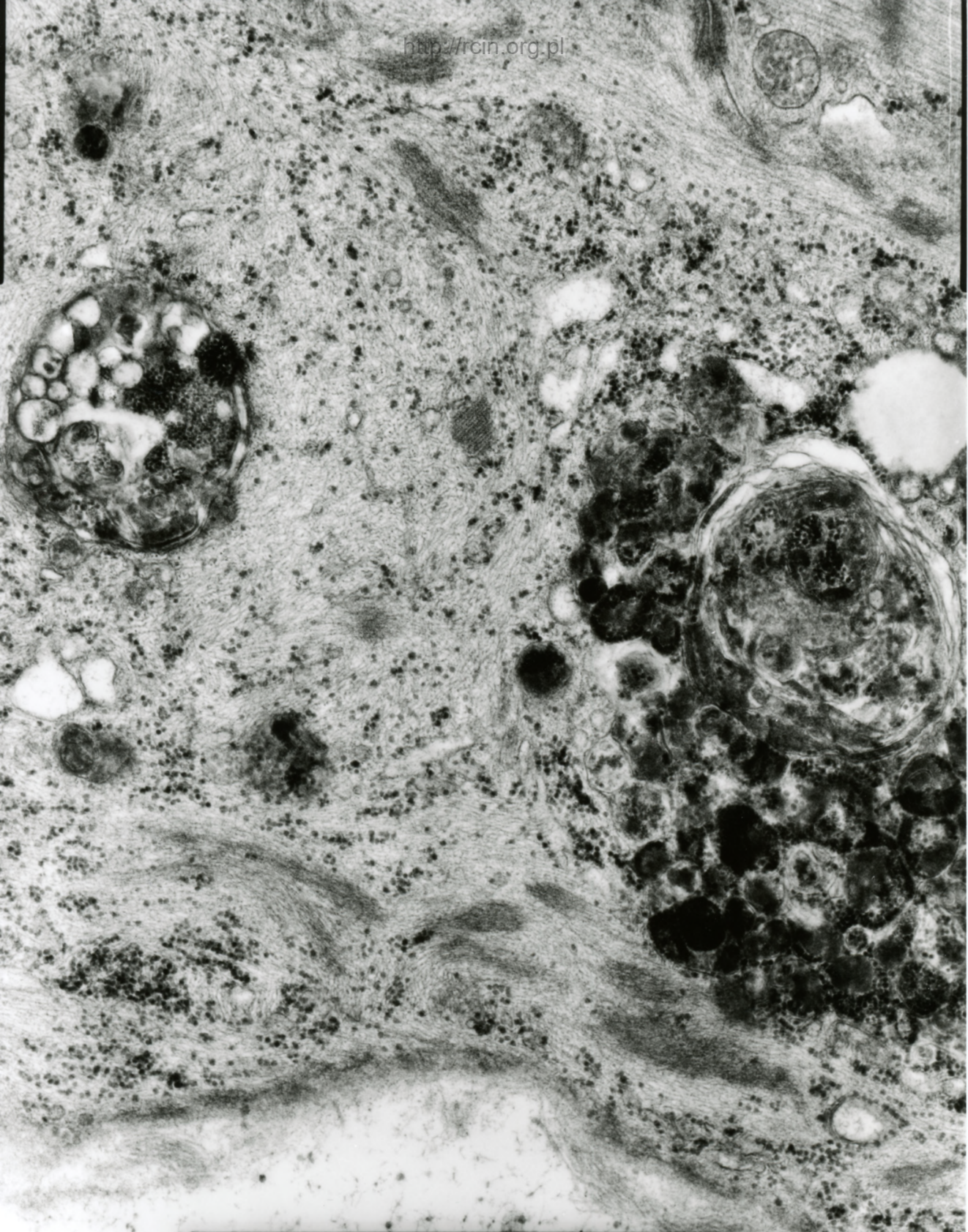
083364

RR OKU

X12K

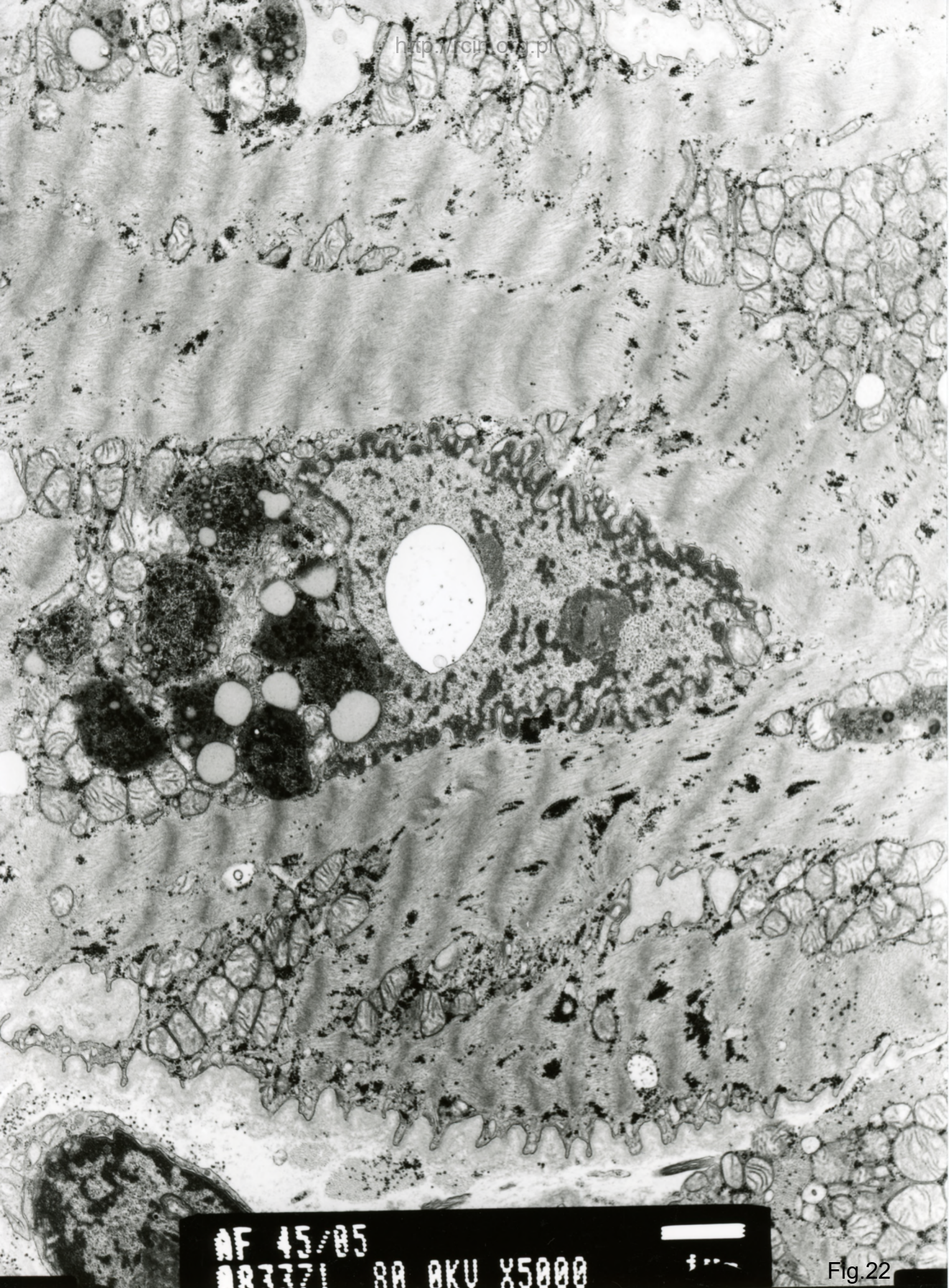
500

Fig.20



AF 45/05/W
060016 80.0KV X15K 500nm

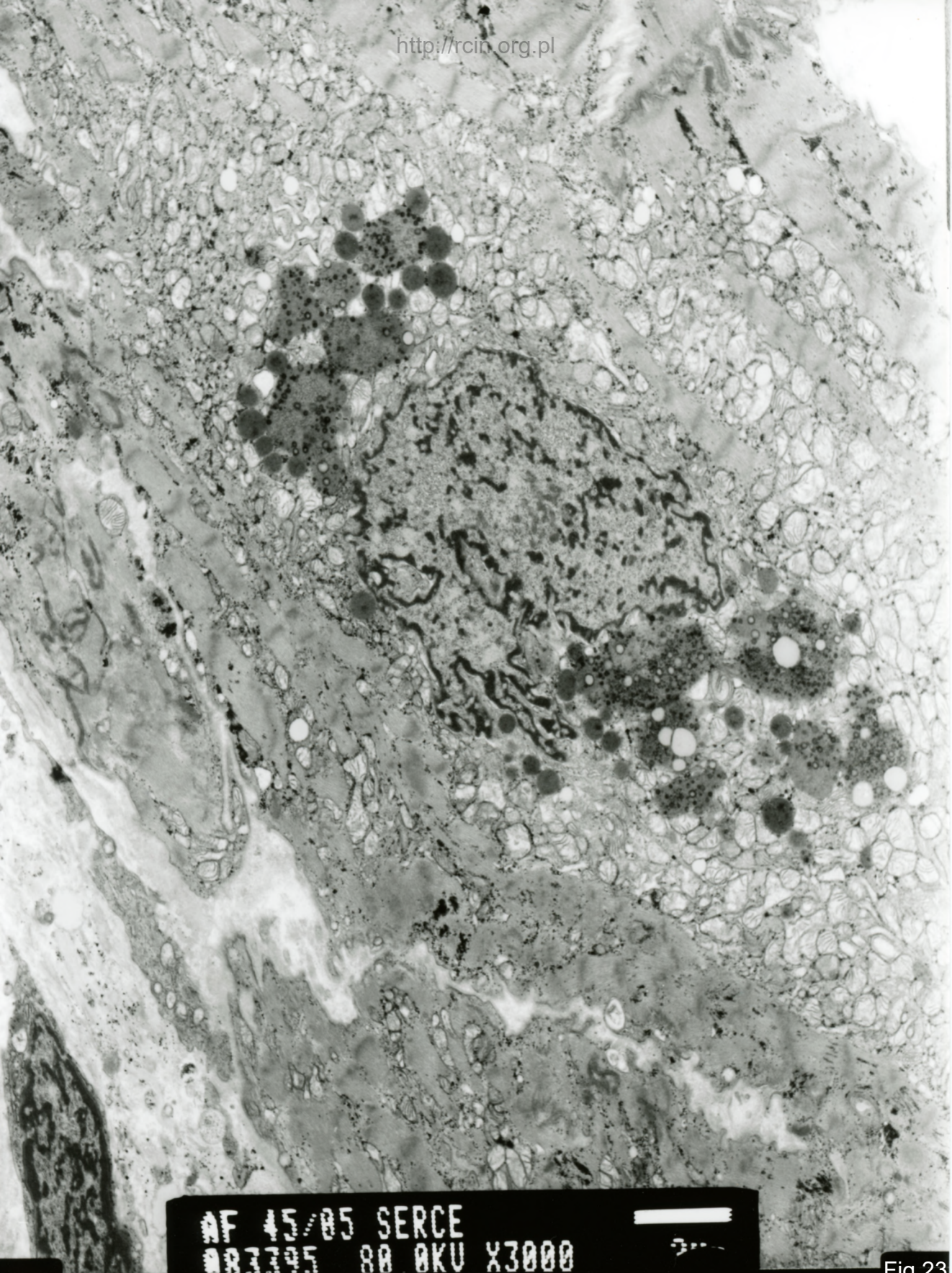
Fig.21



AF 45/05

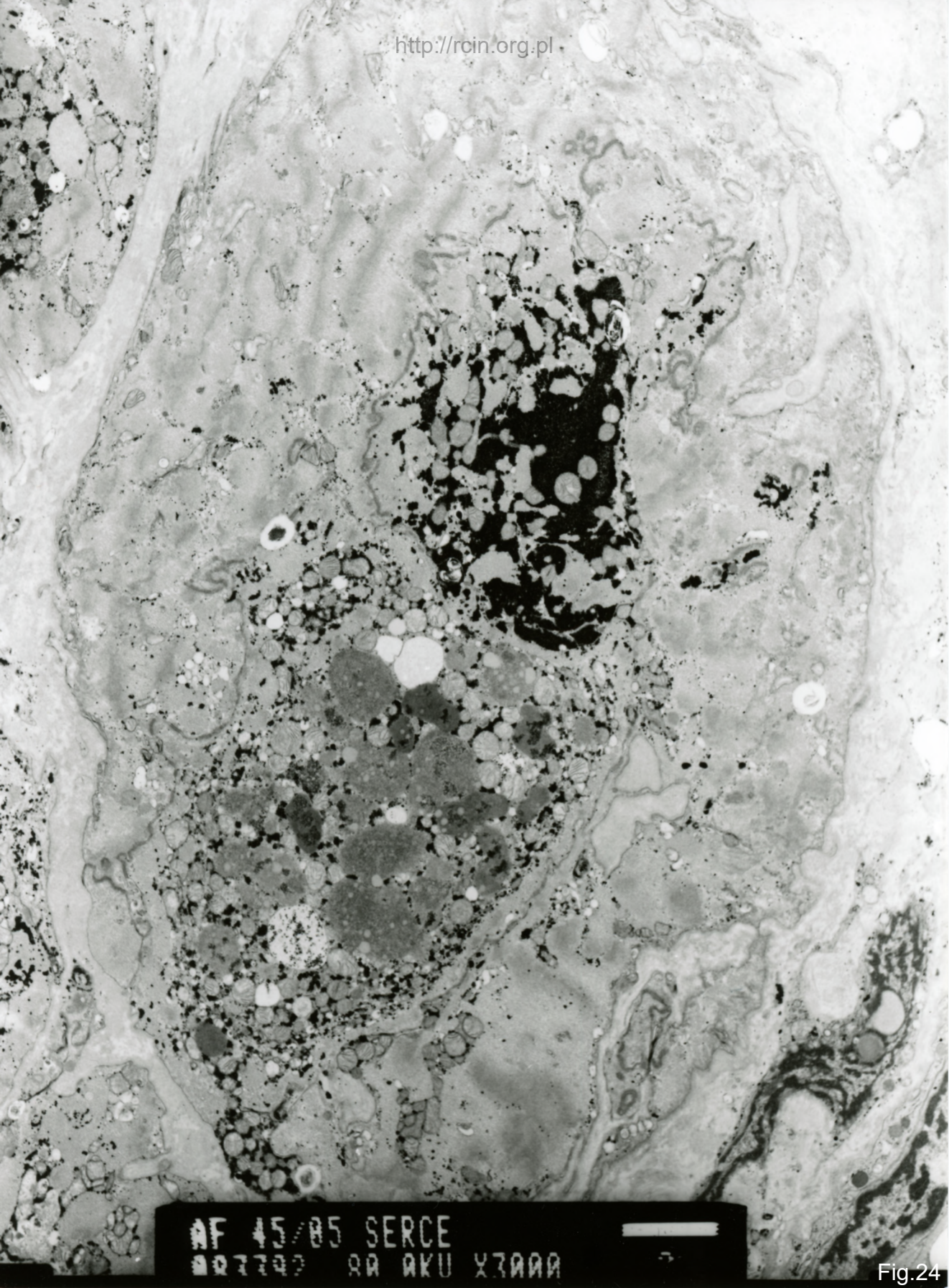
003371 80 0KU X5000

Fig.22



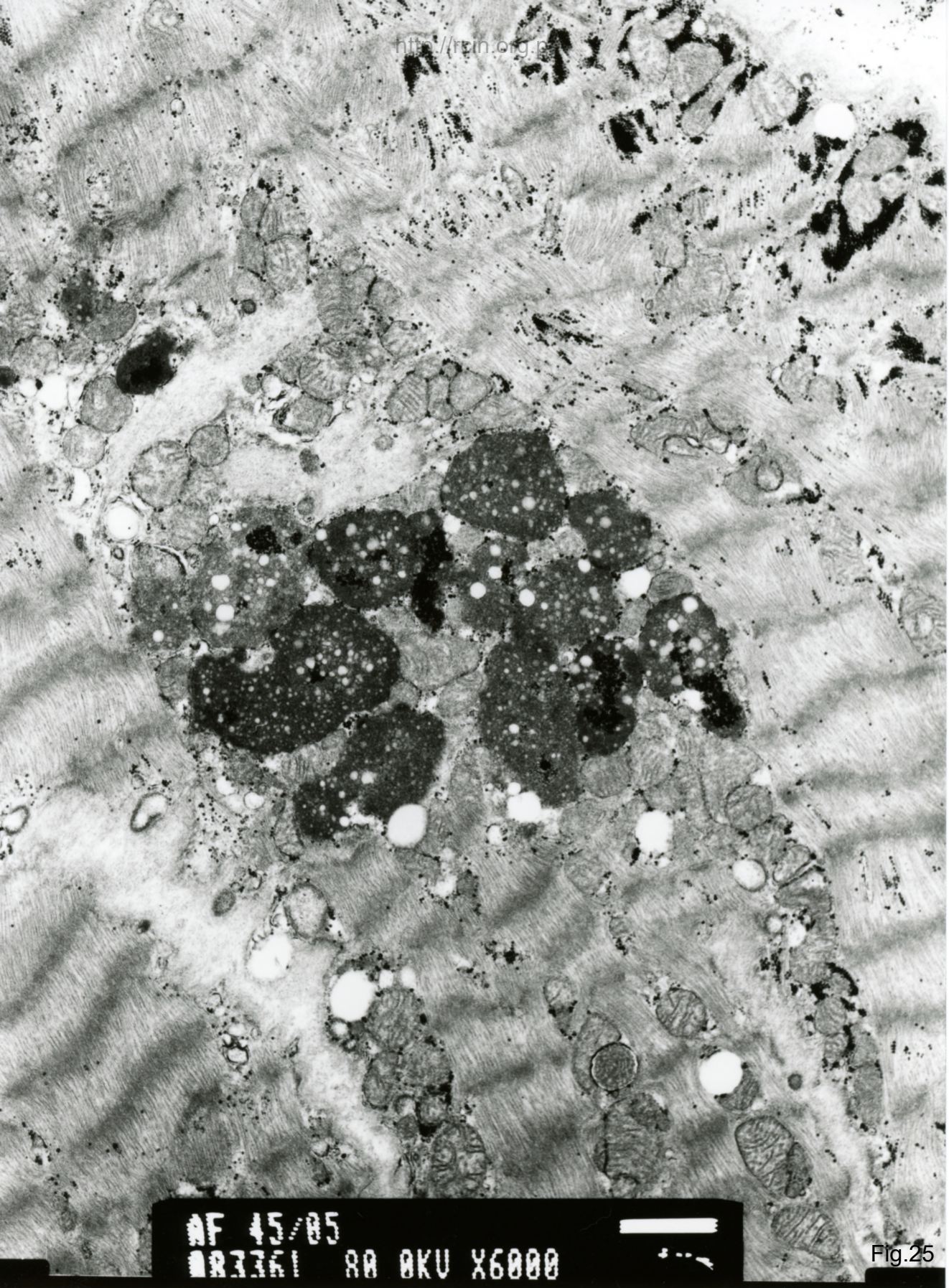
AF 45/05 SERCE
003395 80 0KV X3000

Fig.23



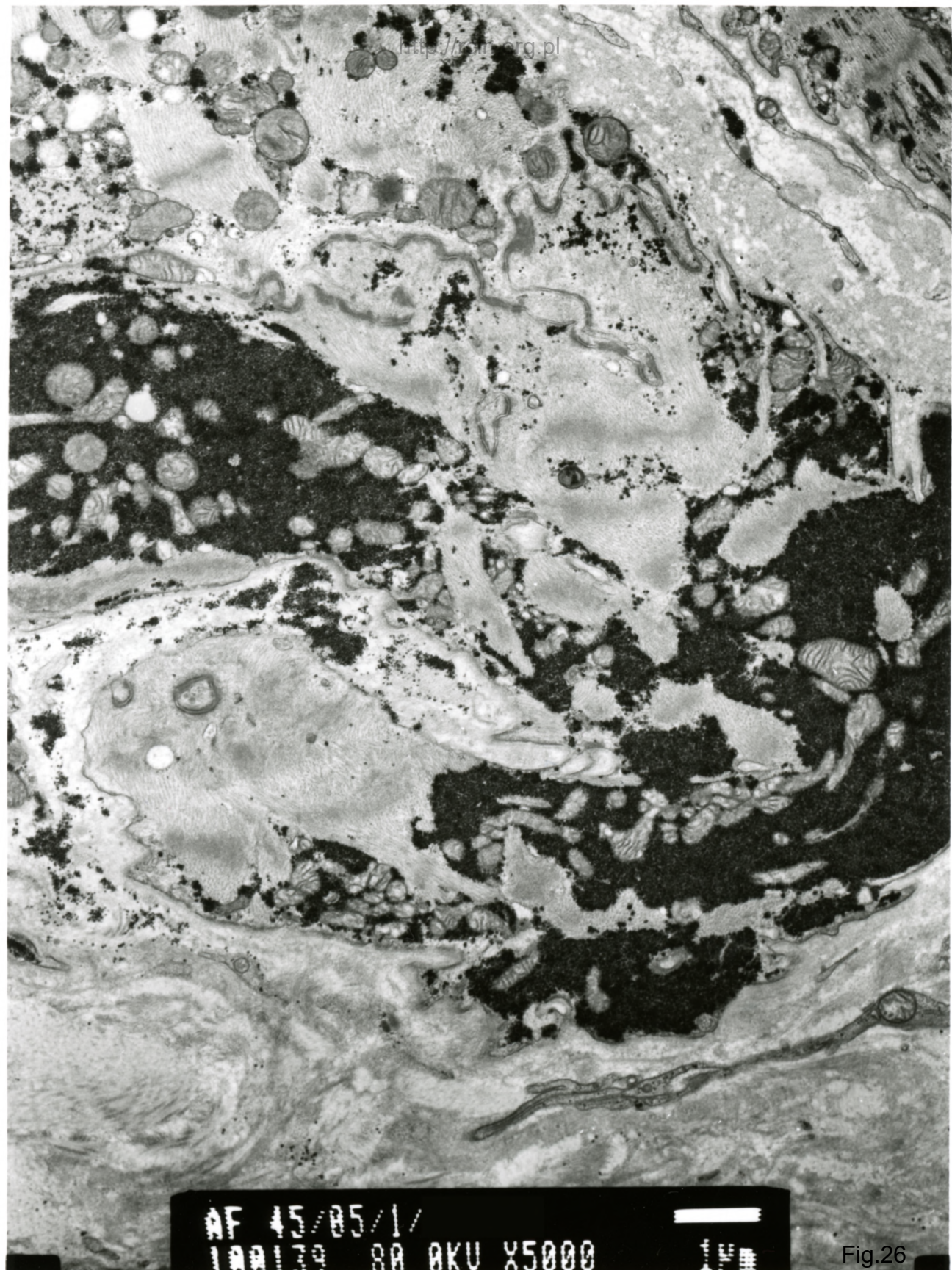
AF 45/85 SERCE
007790 RA AKU X3000

Fig.24

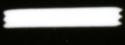


AF 45/05
083361 RR 0KU X6000

Fig.25

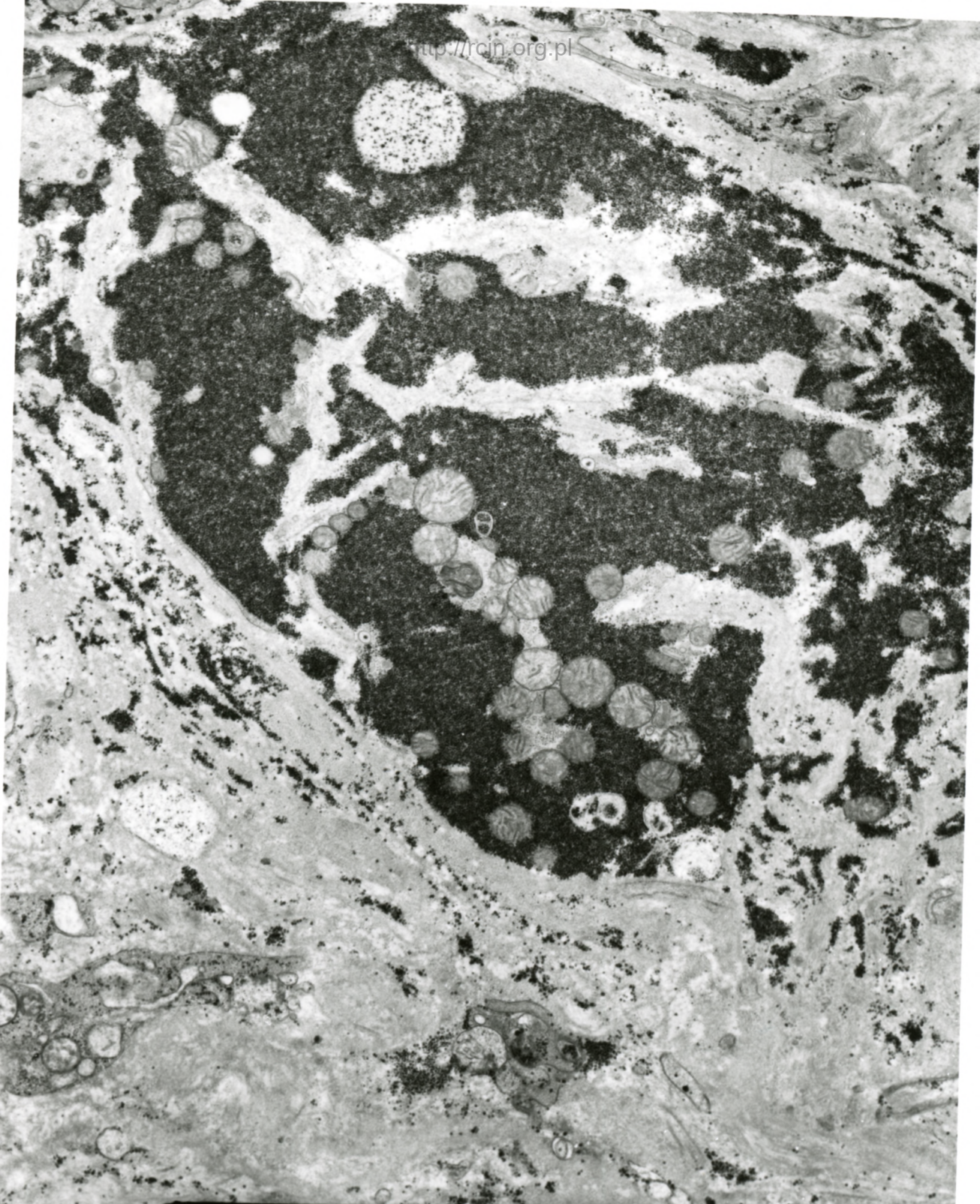


AF 45/05/1/
100139 80.0KV X5000



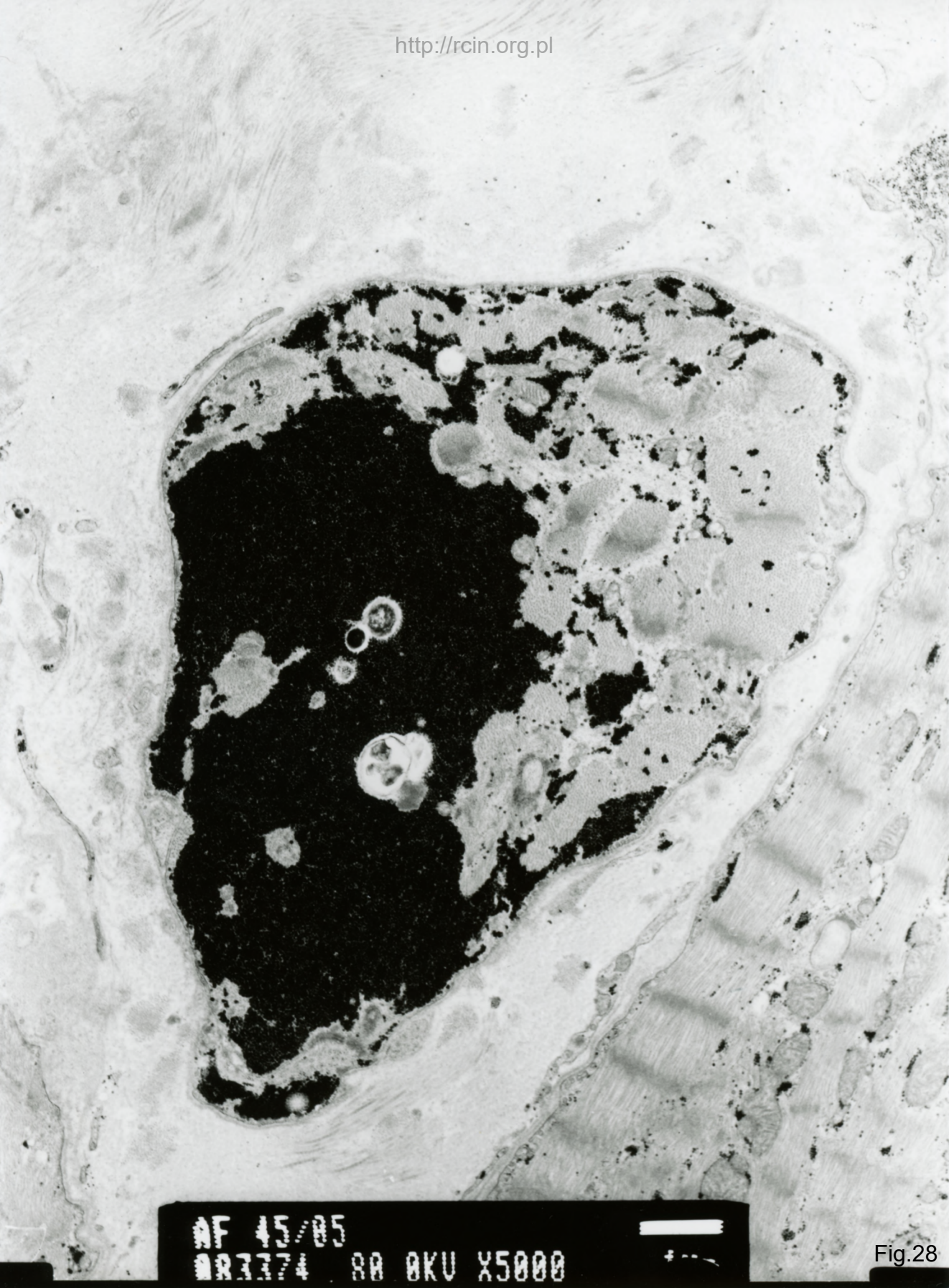
1µm

Fig.26



AF 45/05/1/
100140 80.0KV X5000 1μm

Fig.27



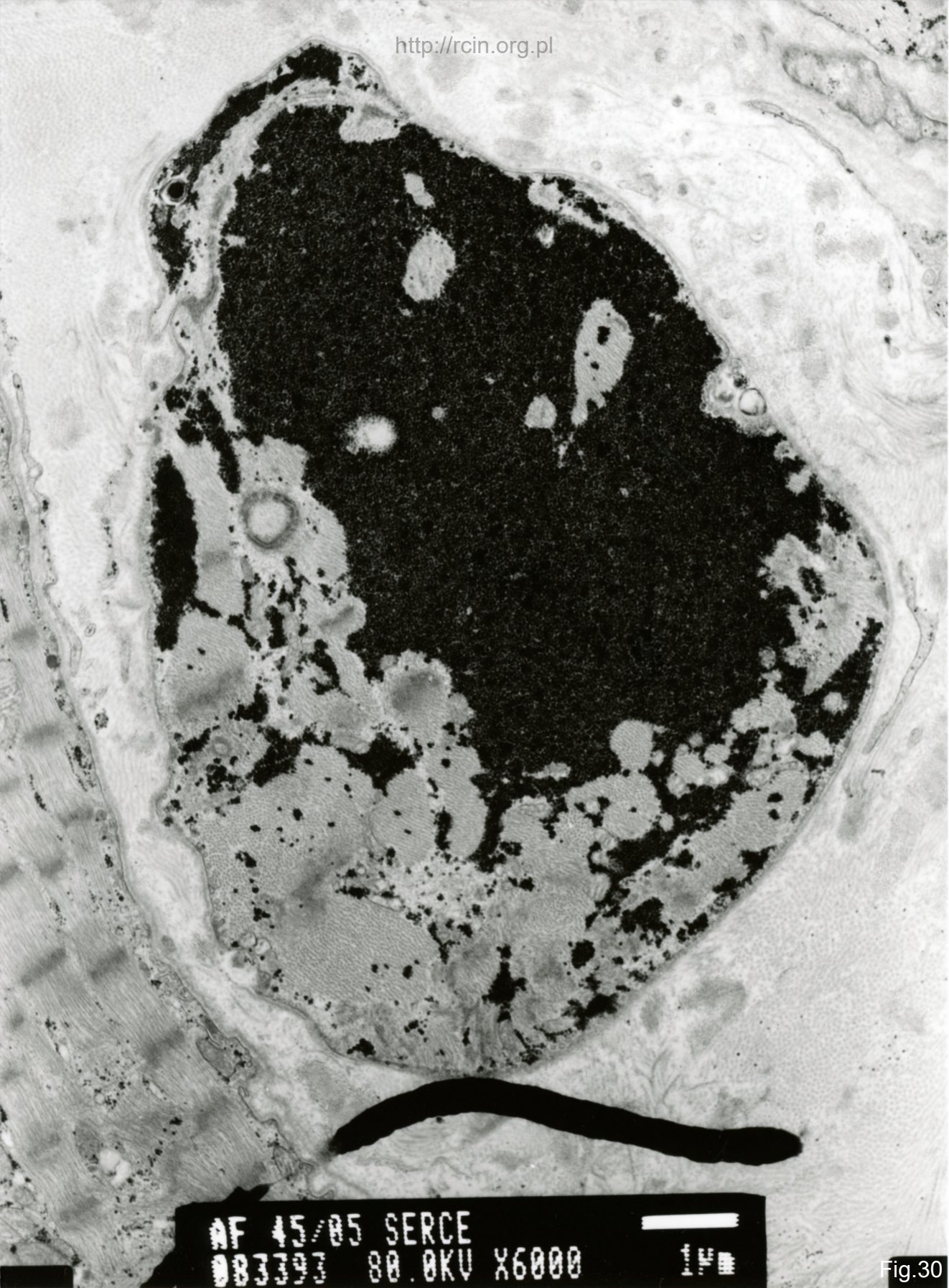
AF 45/85
003374 80 0KU X5000

Fig.28

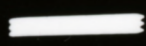


AF 45/05 SERCE
003794 RA OKU X6000

Fig.29

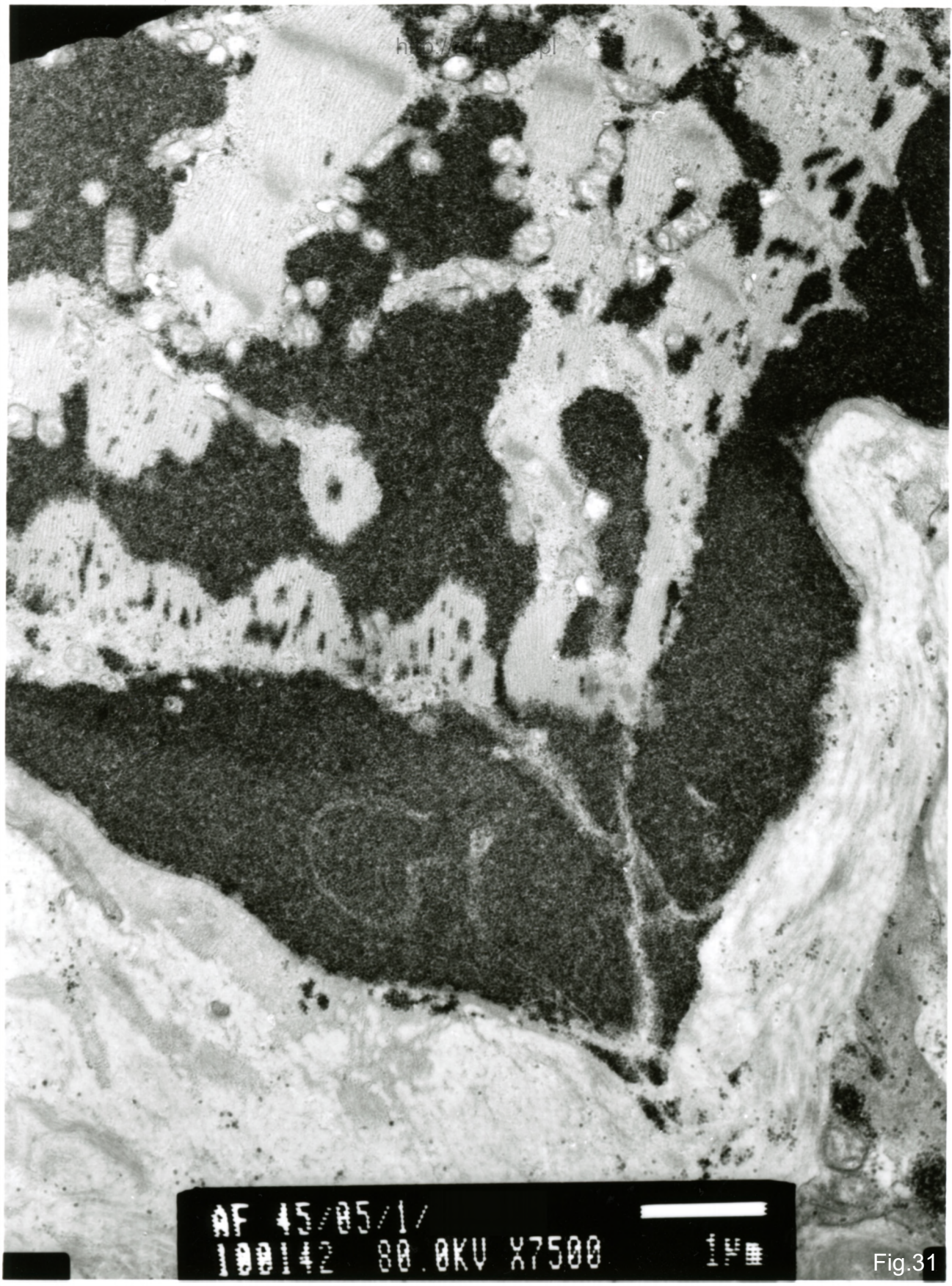


AF 45/85 SERCE
083393 80.0KV X6000



1 μm

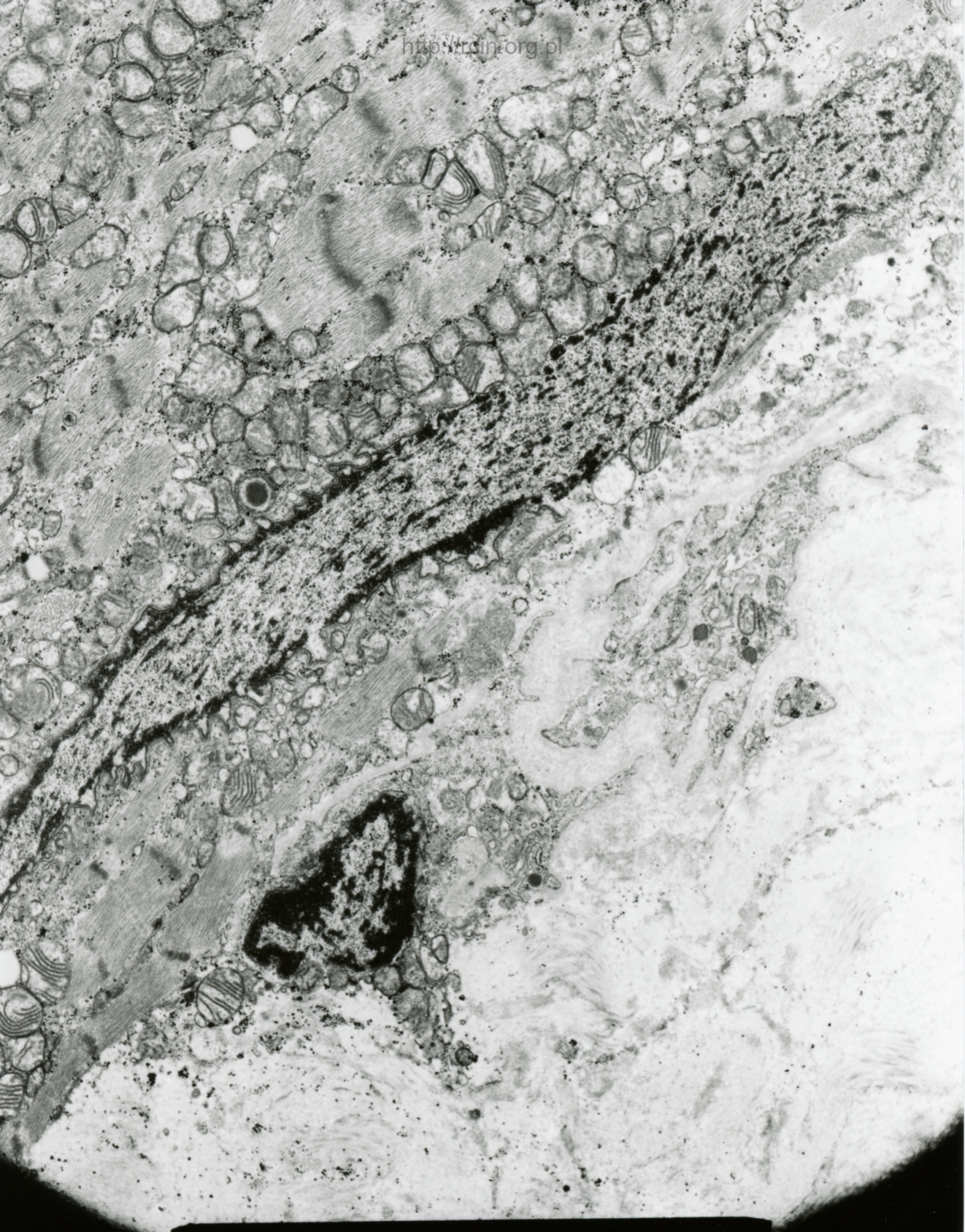
Fig.30



AF 45/85/1/
100142 80.0KV X7500

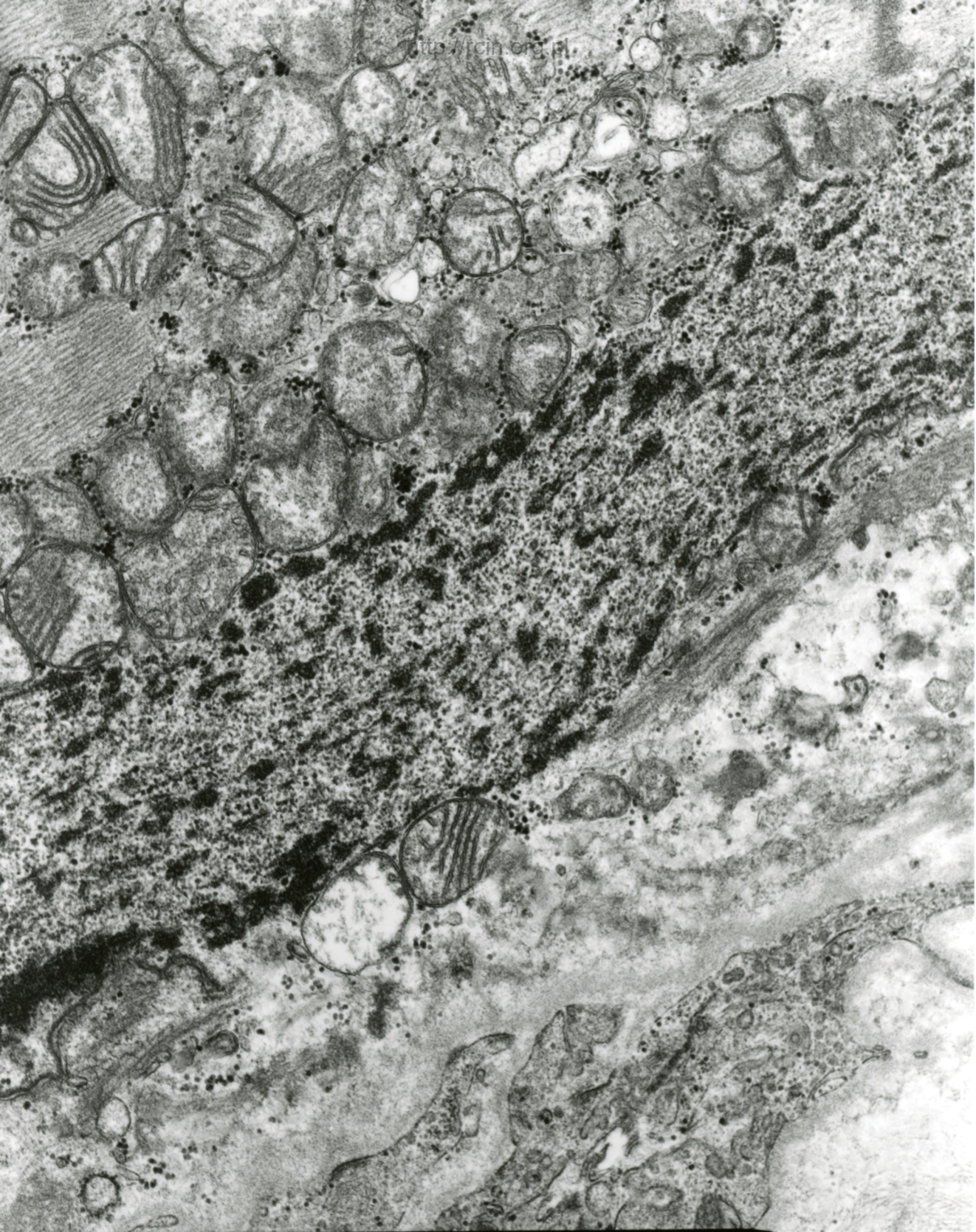


Fig.31



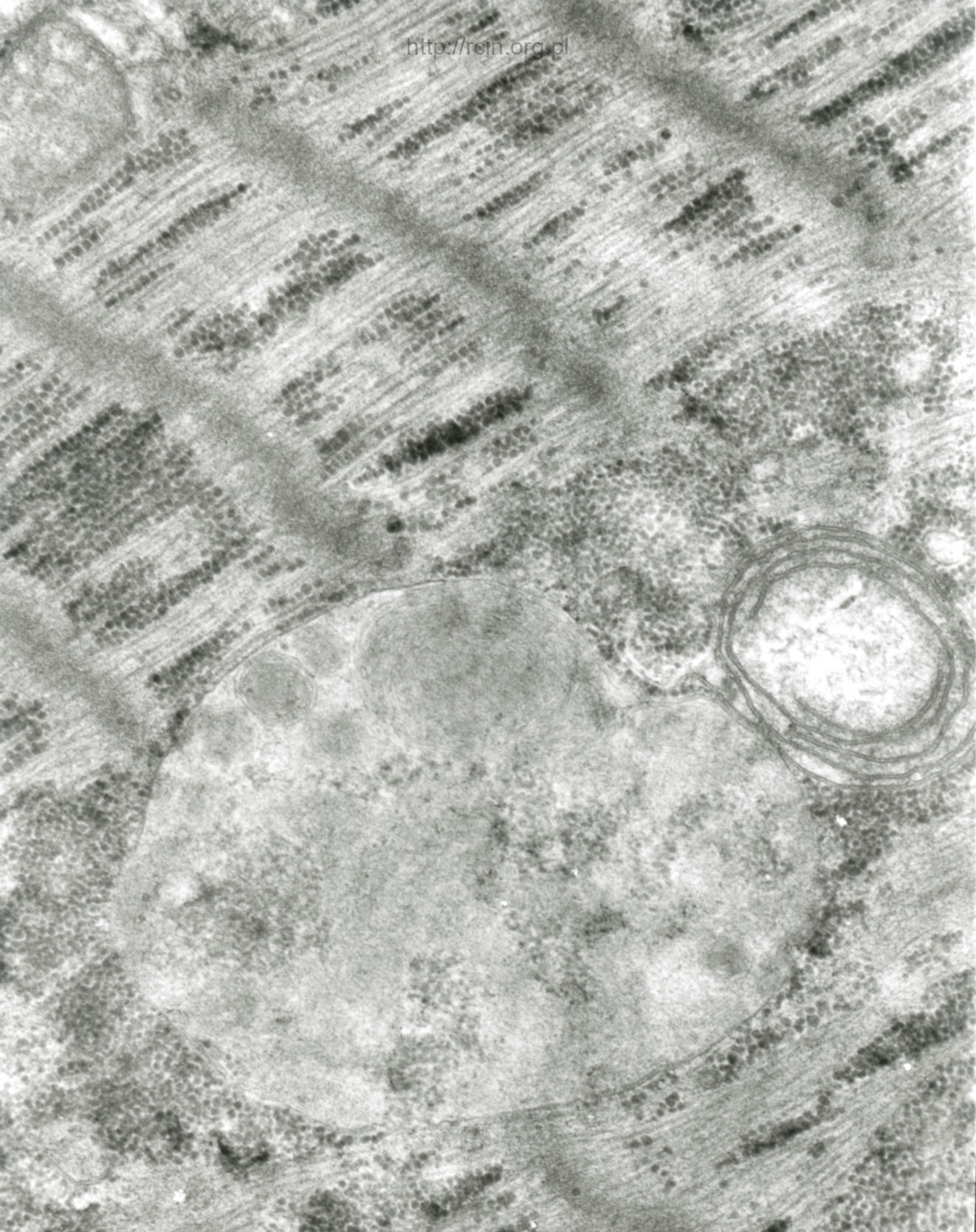
AF 45/85
003365 RA OKU X5000

Fig.32



AF 45/05
083366 RA OKU X12K 500

Fig.33



AF 45/05/SERCR
050503 80.0KV X20K 200nm

Fig.34

45/05

1. 32

Podejrzenie amyloidozy/kardiomiopatii mitochondrialnej/zapalenia mięśnia sercowego

We fragmentach biopsjatu widoczne zachowane miofibrylle i niezmienny układ sarkomerów obserwuje się jednak znaczną nukleopatię – jądra kardiomiocytów o nieprawidłowym kształcie, pofałdowanej otoczce a niektóre zawierające w swym wnętrzu organelle komórkowe (Fig. 1-8). Ocena ultrastrukturalna wykazała na pewnych obszarach zaburzenia w układzie sarkomerowym i w przebiegu miofilamentów oraz ich dezorganizację (Fig. 9,10,11). Obserwowano licznie występujące struktury autofagalne (Fig. 12-21) oraz złogi lipofuscyny, obecne zwłaszcza w pobliżu jąder kardiomiocytów (Fig. 22-25). W biopsjacie widoczne były fragmenty tkanki o znacznym nagromadzeniu glikogenu (Fig. 26-31). Niektóre mitochondria charakteryzowały się jasną macierzą mitochondrialną i ubytkiem grzebieni bądź ich nieprawidłowym przebiegiem (Fig. 32,33,34).

Suspected amyloidosis / mitochondrial cardiomyopathy / myocarditis

Electronmicroscopy evaluation revealed in some parts of the biopsy the presence of preserved myofibrils and an unchanged sarcomeres pattern, however, a significant nucleopathy was observed. Cardiomyocyte nuclei were characterized by abnormal shape and some of them by discontinuity of the nuclear membrane and contained cellular organelles inside (Figs. 1-8). Focal changes in the course of myofilaments and their disorganization as well as changes in sarcomere pattern were observed (Figs. 9,10,11). Numerous autophagous structures were seen (Figs. 12-21) Also some lipofuscin deposits, especially in the vicinity of cardiomyocyte nuclei were noticed (Figs. 22-25). The biopsy analyses showed tissue fragments with significant glycogen accumulation (Figs. 26-31). Some mitochondria were characterized by a bright mitochondrial matrix and a loss or abnormal course of mitochondrial cristae (Figs. 32,33,34).