

Badanie wycinka mięśniowego w mikroskopie świetlnym

Nr: 10/02

Imię i nazwisko:

Wiek : 12

Rozpoznanie : DMP ; Emery-Dreyfuss

Mięsień : dwugłowy ramienia lewego

Data pobrania : 23.01.02

Włókna przeroste o średnicy prawidłowej i bardzo małe wymieszane nieregularnie tworzą pęczki oddzielone znaczną ilością tkanki łącznej. Szereg włókien ulega rozszczepieniu. Widoczne są pojedyncze włókna uległe martwicy, pojedyncze regenerują. Ponadto w niektórych włóknach widoczne są wodniczki różnego kształtu i wielkości czasem szczelinowate odpowiadające morfologia „rimmed vacuoles” . Znaczny rozrost tkanki łącznej w endomysium.

Podział włókien na typy metaboliczne zachowany. We włóknach typu 1 obserwuje się drobne ubytki aktywności okrągłe, regularne podobne mini-core.

W sumie cechy uszkodzenia pierwotnie mięśniowego dość dużego stopnia z obecnością „ rimmed vacuoles” . Konieczna kontrola M-E, podejrzenie wtrętów YBM podobnych.

Prof. dr hab. Anna Fidziańska-Dolot

10/02

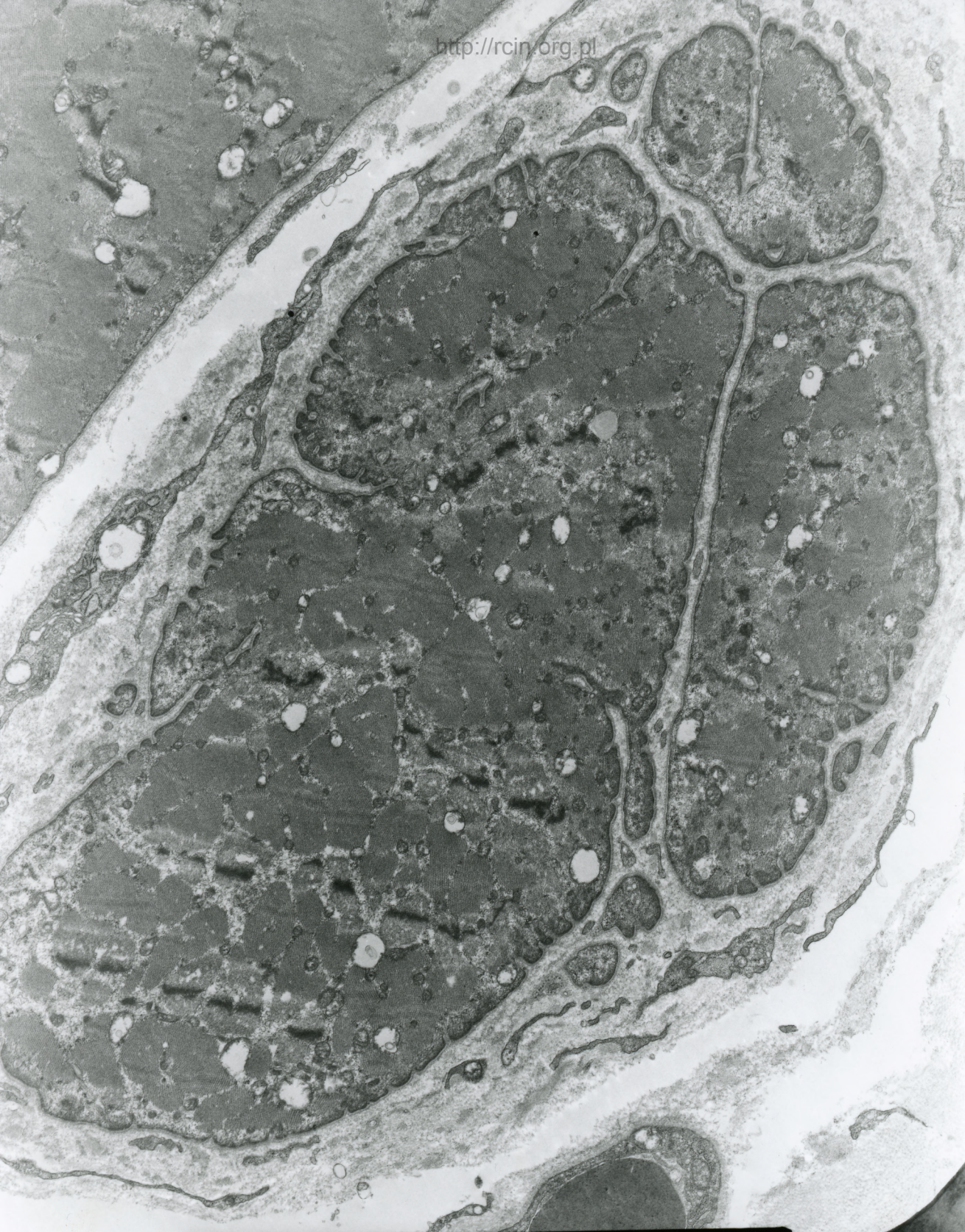
Rozpoznanie: podejrzenie DMP, dystrofii mięśniowa Emery'ego-Dreyfussa

Analiza elektronowo-mikroskopowa wykazała znaczne zmiany w obrębie miocytów. Obserwowano zaburzenia w układzie sarkomerów i rozproszenie miofibryli (Fig. 1-6). Bardzo licznie występowały wakuole autofagalne wypełnione strukturami mielinopodobnymi i debrisy komórkowym (Fig. 7-21). Miejscami, w pobliżu wakuoli, widoczne były inkluzje włóknikowego materiału (Fig. 22). Obserwowano znaczne zmiany ultrastrukturalne w jądrach komórek mięśniowych. Widoczny był ubytek chromatyny, zwłaszcza w części środkowej jąder i pęcherzyki (Fig. 23-27), oraz obecność struktur mielinopodobnych i wakuoli (Fig. 28-33) w ich wnętrzu. W obrębie części jąder komórek mięśniowych widoczne były struktury włóknikowe (Fig. 34-41) w innych tubularne (Fig. 42-46). Włókna mięśniowe oddzielone znaczną ilością tkanki łącznej (Fig. 47-50).

The patient with suspected DMP and Emery-Dreyfuss muscular dystrophy was examined. A left biceps muscle biopsy was taken.

Electron-microscopic analysis revealed significant changes within the myocytes. Changes in sarcomere pattern were observed. The myofibrils were disorganized and scattered (Figs. 1-6). Autophagic vacuoles filled with myelin-like structures and cellular debris were abundant (Figs. 7-21). Focally, in the vicinity of vacuoles, inclusions of filamentous material were visible (Fig. 22). Substantial ultrastructural changes were observed in muscle cell nuclei. Chromatin loss, especially in the central part of the nuclei and heterochromatin blebs formation were seen (Figs. 23-27). The presence of myelin-like structures and vacuoles (Figs. 28-33) inside the nuclei were noticed. Within some of the muscle cell nuclei filamentous structures (Figs. 34-41) in others rod-like structures (Fig. 42-46) were visible. Muscle fibers were separated by abundant connective tissue (Figs. 47-50).

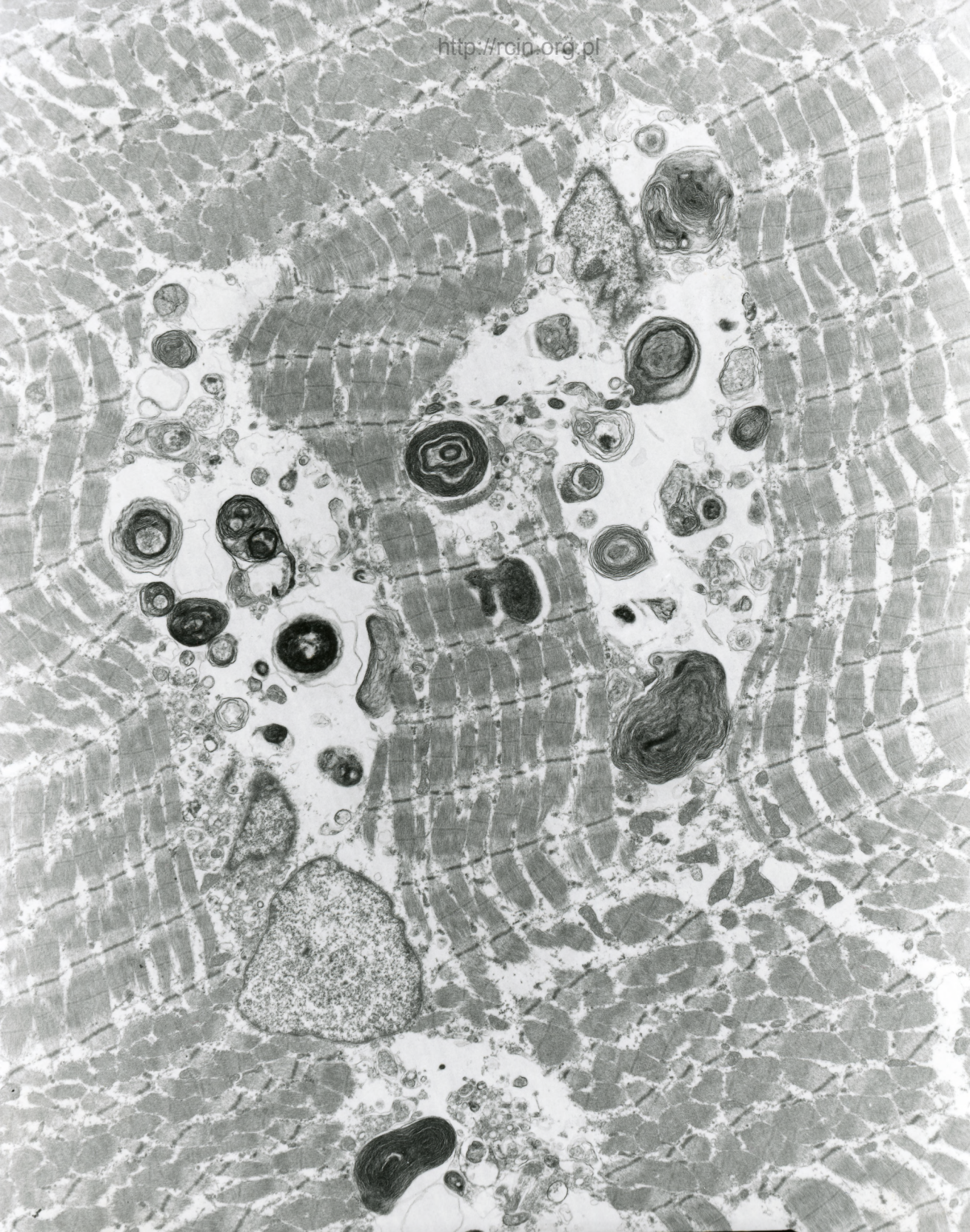




AF 10/02/01  
225502 80.0KV X3000 ZF

Fig. 1





AF 10/02/  
2R5640 80.0KV 1X2000 2PB

Fig. 2





AF 10/02/1/ 2R5662 30.0KV X4000 2P

Fig. 3



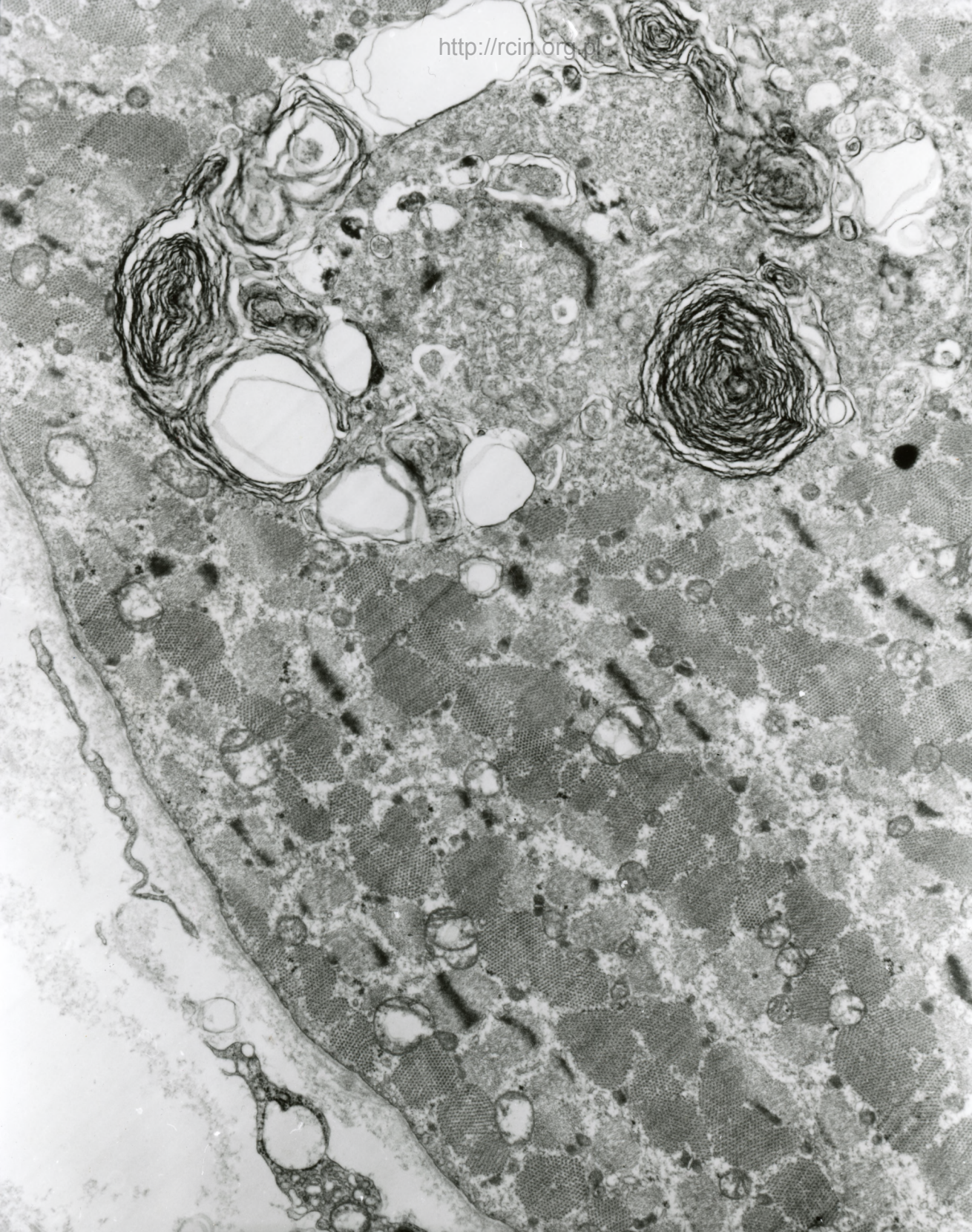


AF 10/02/  
2R5607 00.0KV X5000

IFD

Fig. 4

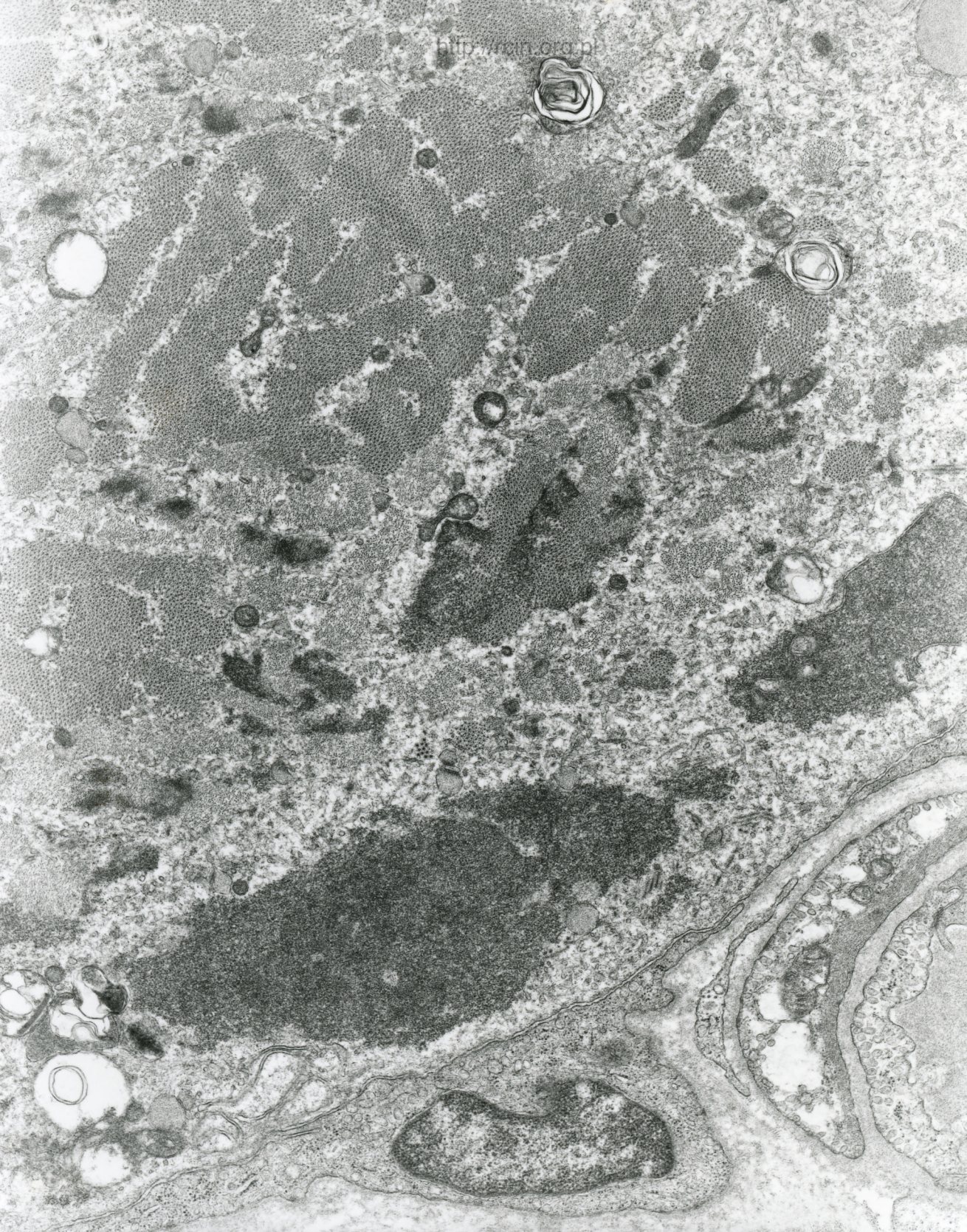




EM 110/02/R  
000000 00.0KV X6000

Fig. 5

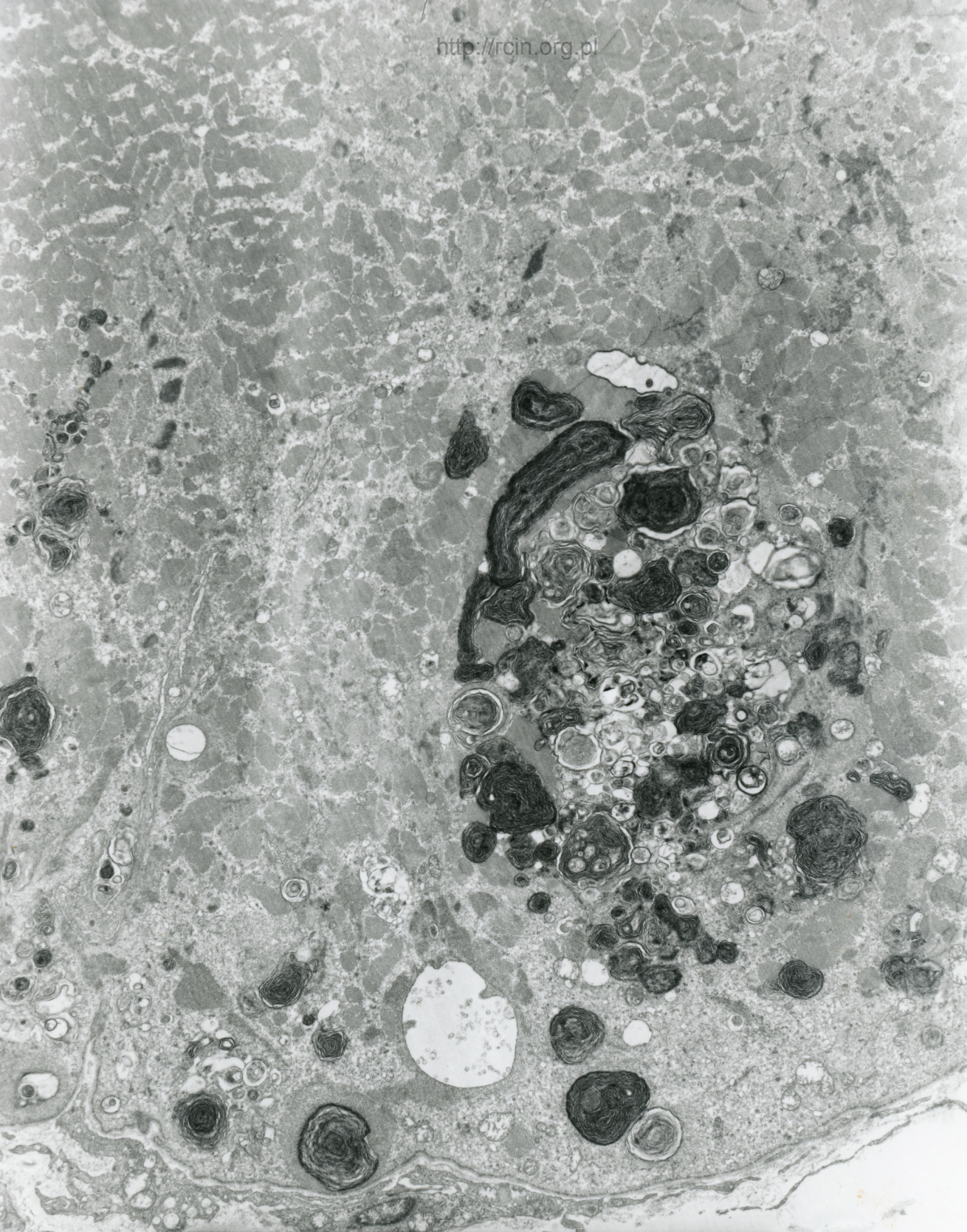




AF 10/02/h/  
2P5862 80 AKU 1X6000 1P

Fig. 6

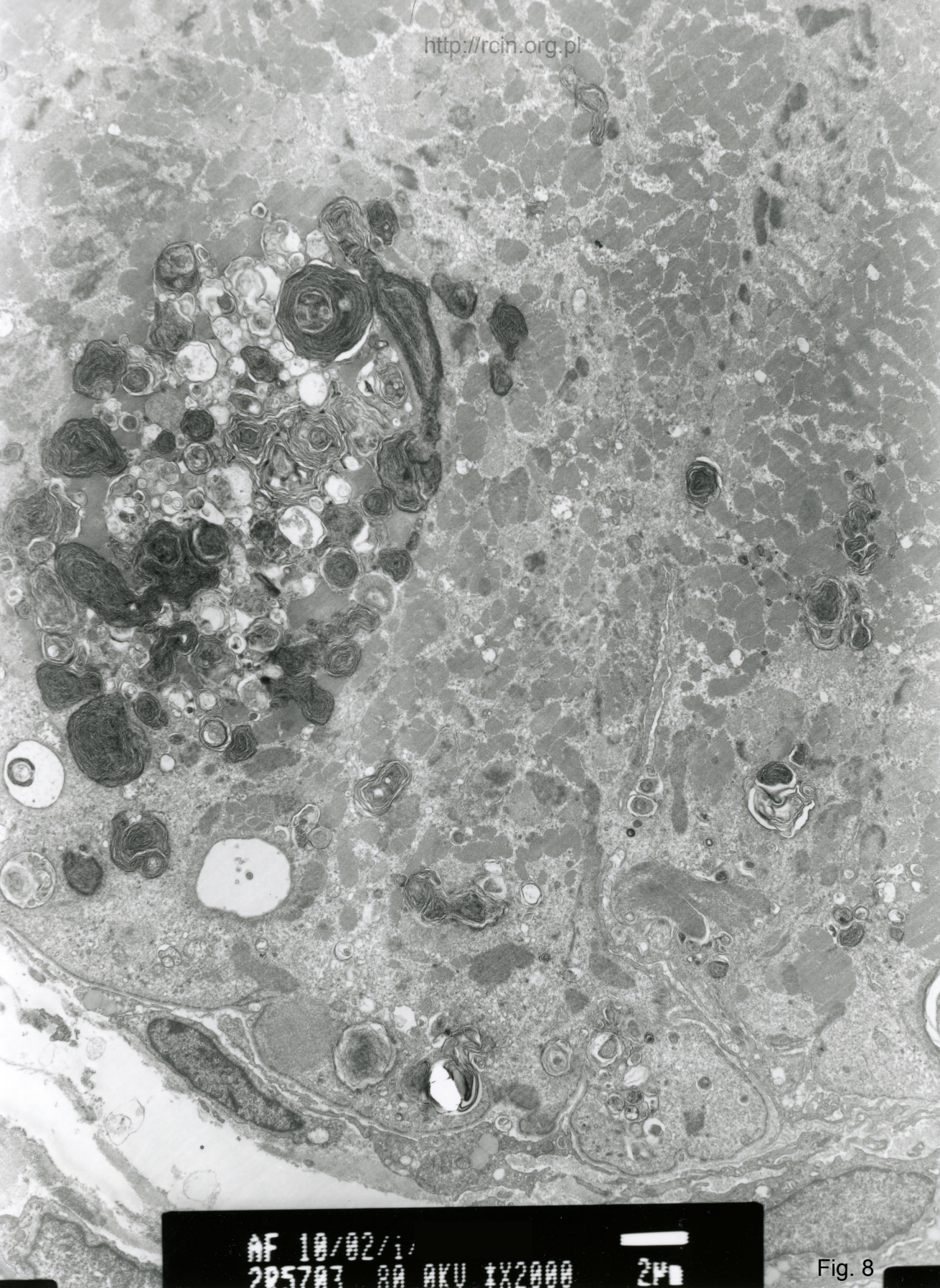




AF 10/02/1/  
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Fig. 7



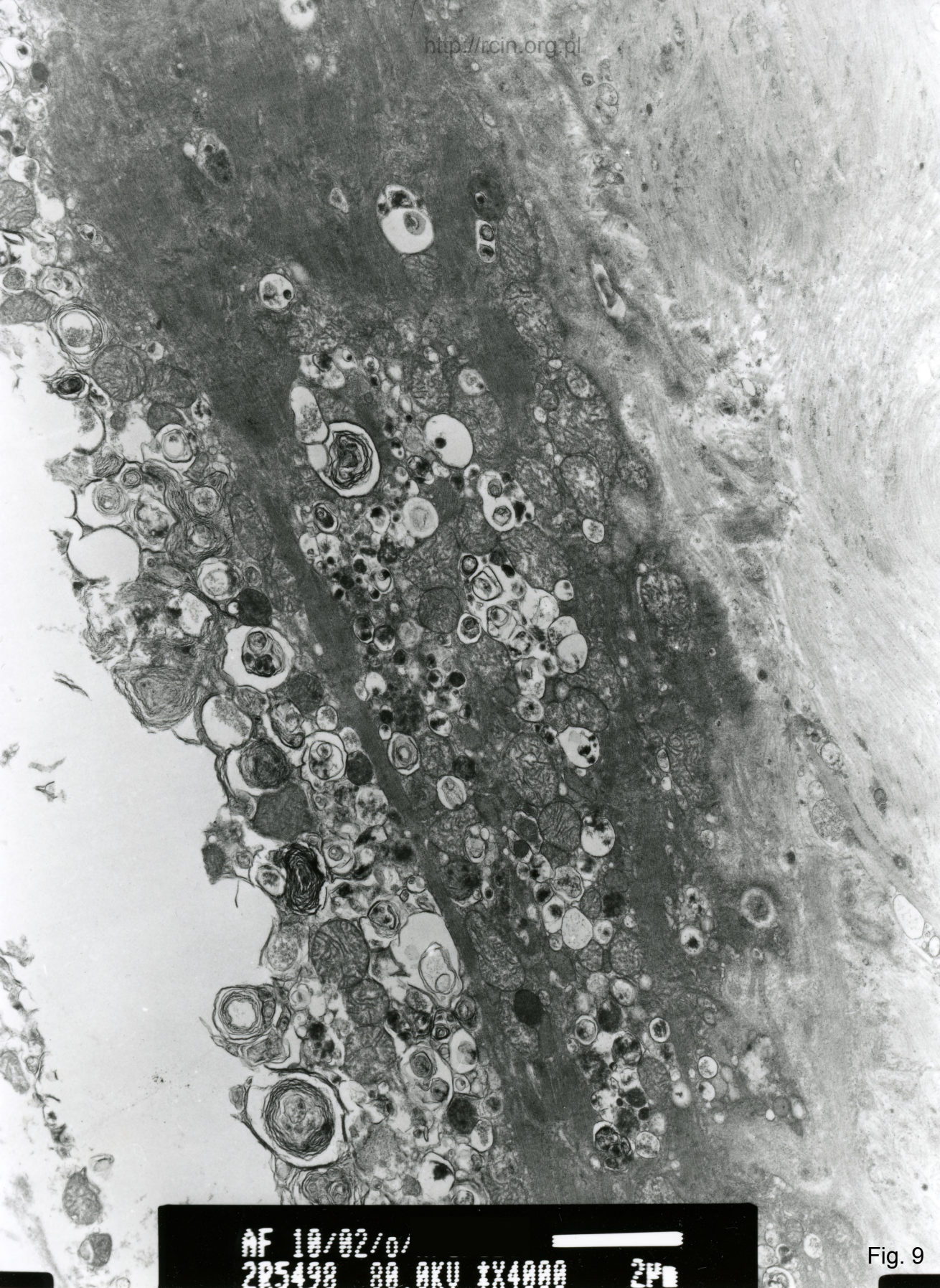


AF 10/02/1/  
205707 00 AKU 1X2000

2µm

Fig. 8

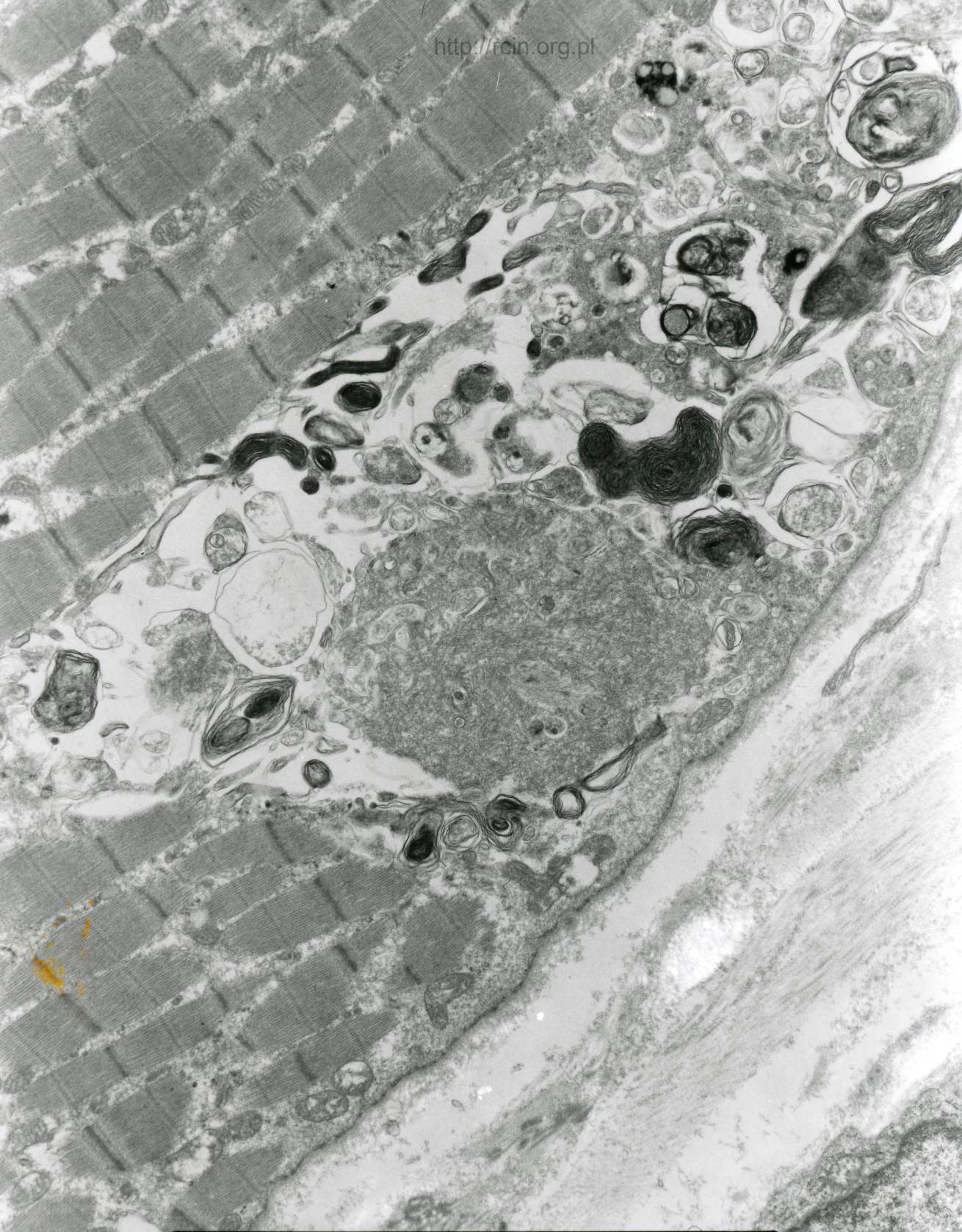




AF 10/02/0/  
205498 80 OKU 1X4000 ZP

Fig. 9

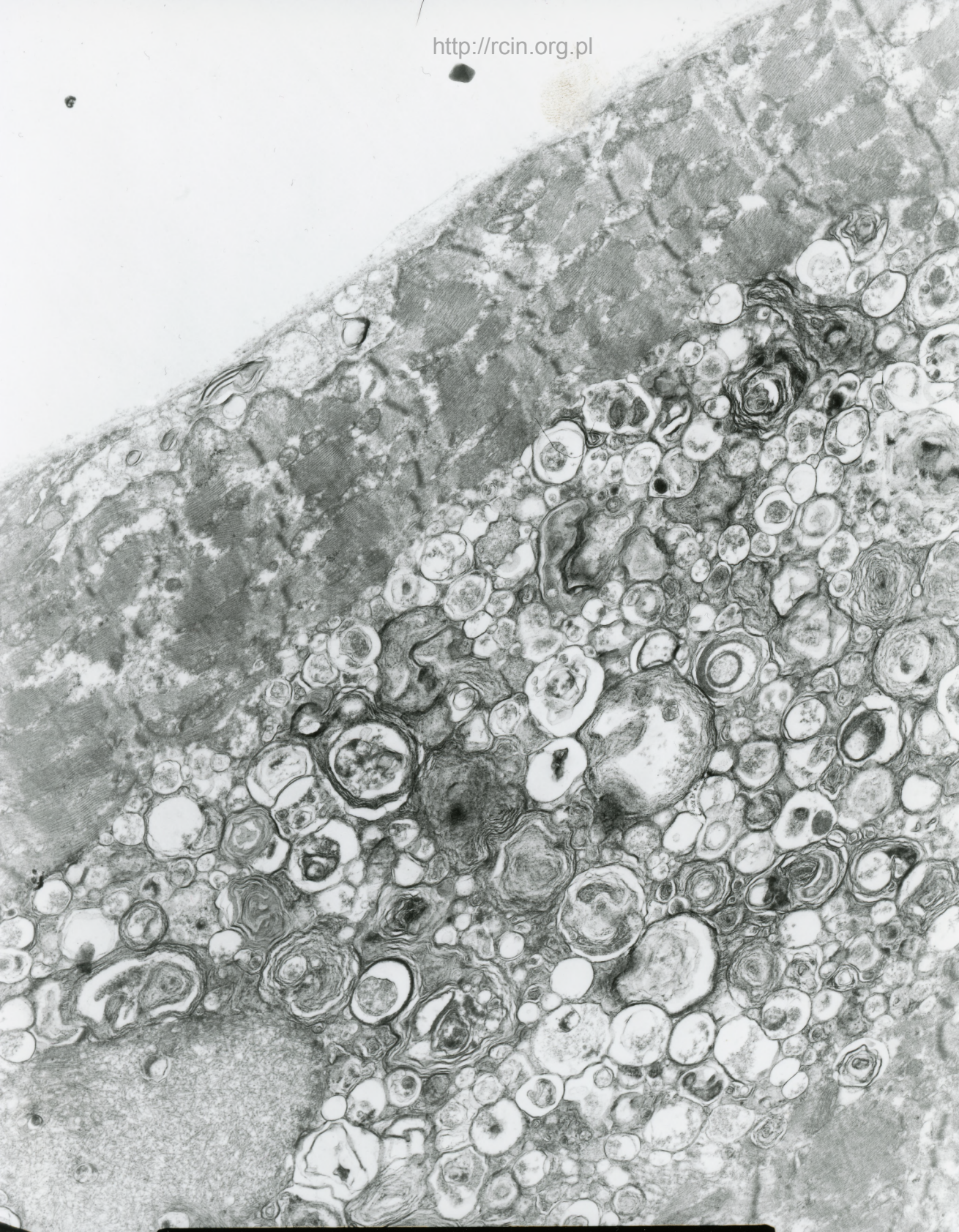




AF 10/02/  
2P5600 SA BKU IX4000 2P

Fig.10

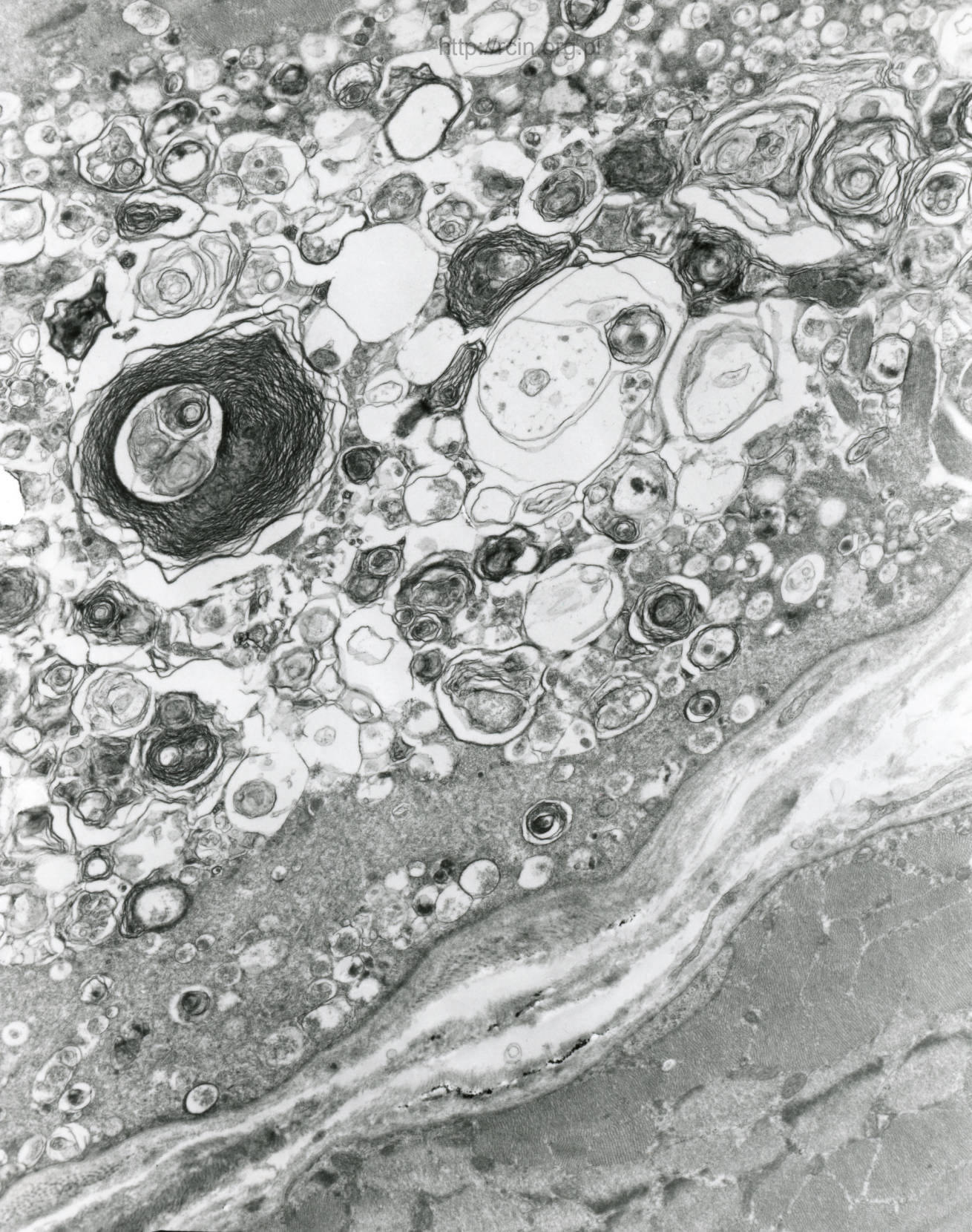




AF 10/02/r/  
205594 80.0KV 1X4000 ZPT

Fig. 11

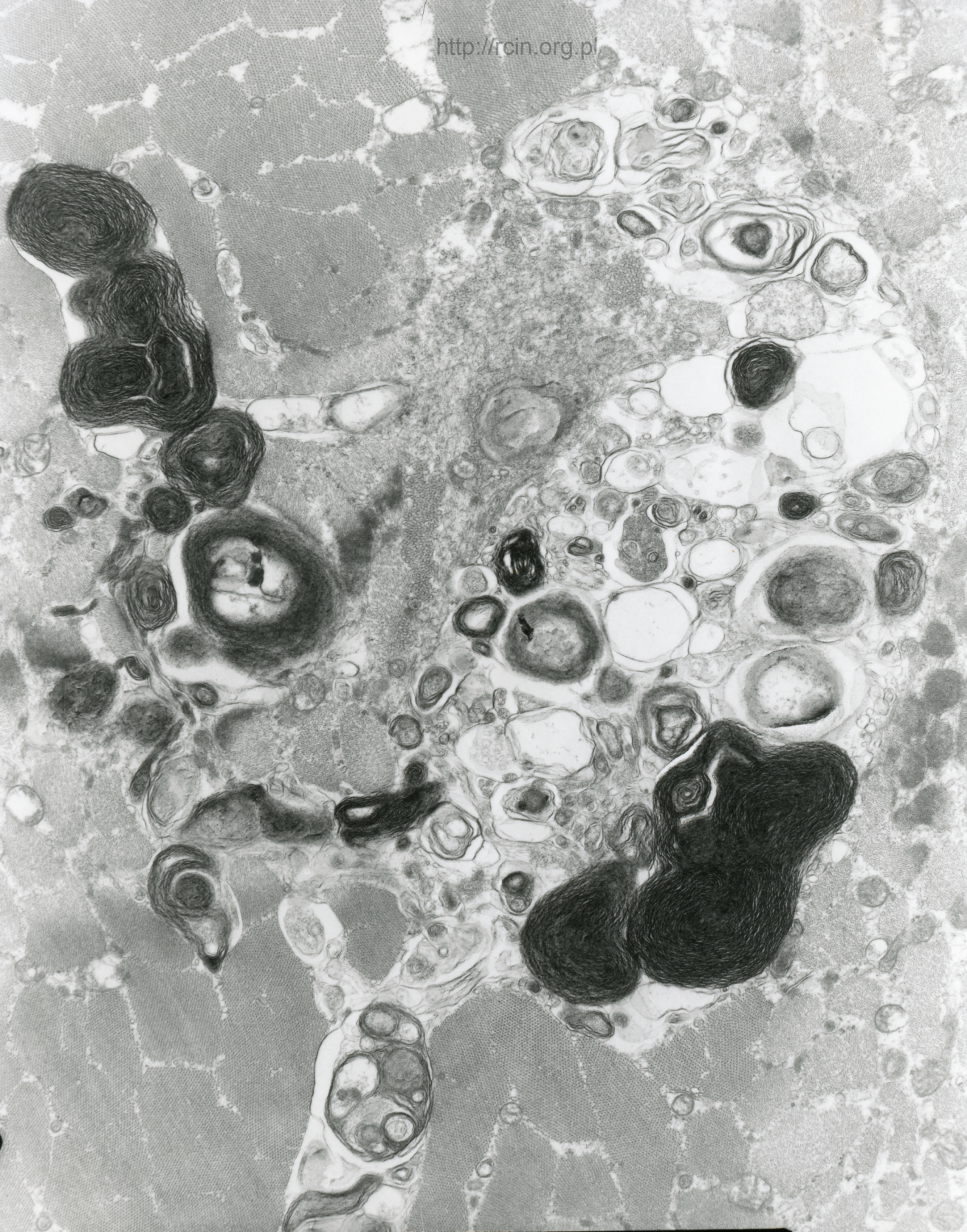




AF 10/02/i  
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Fig. 12

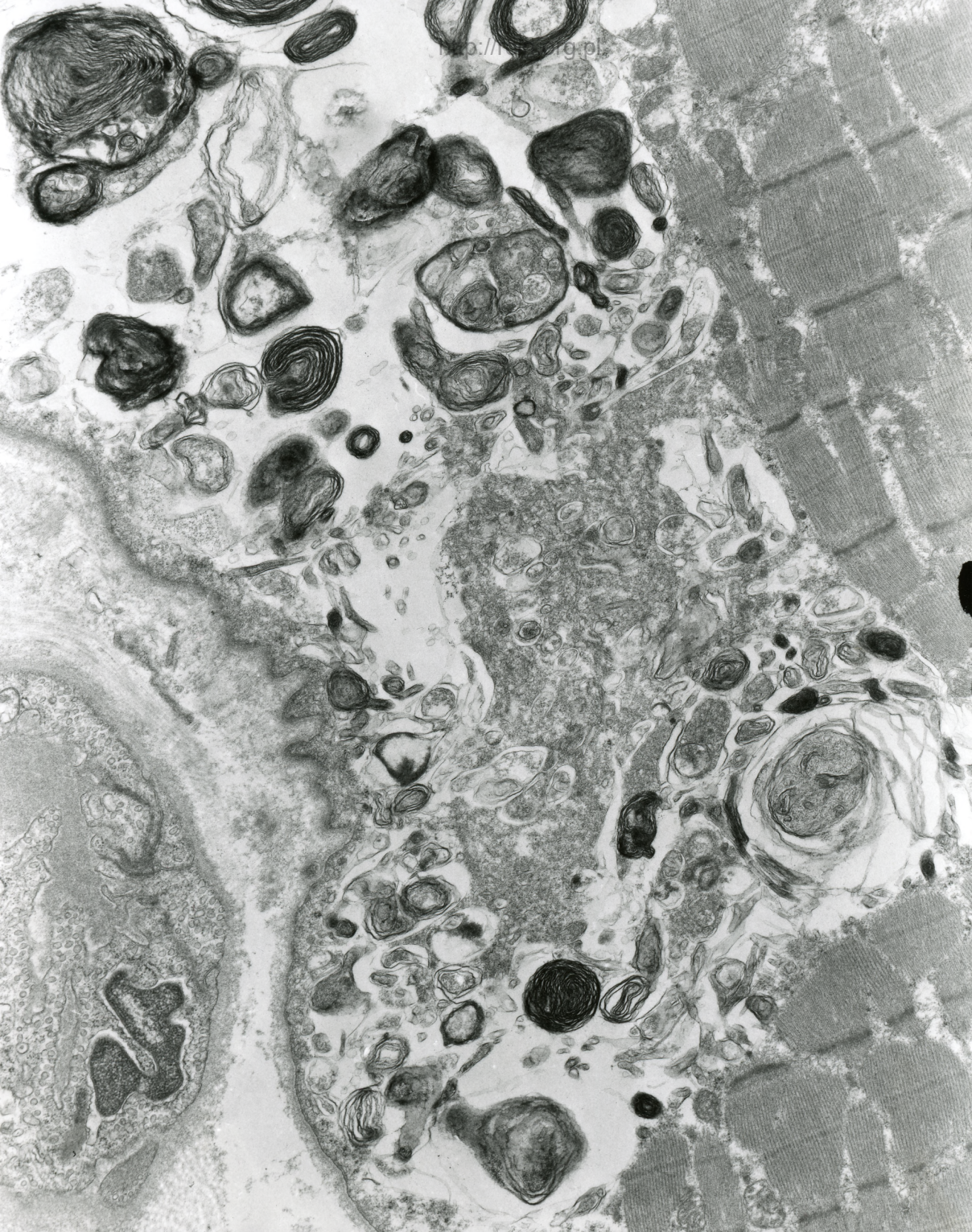




AF 10/02/1/  
2R5659 80.0KV  $\times$ 5000 1 $\mu$ m

Fig. 13

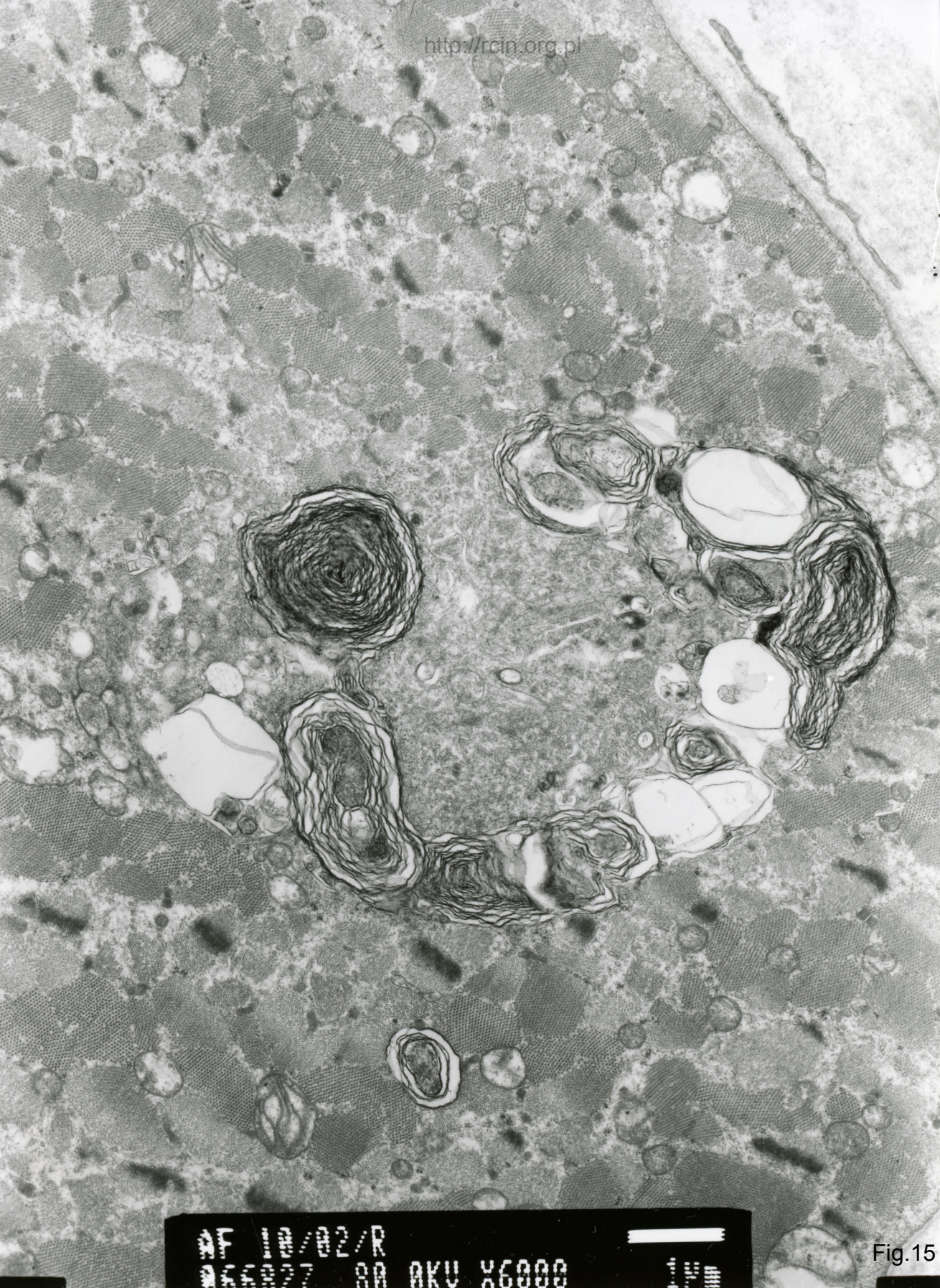




AF 10/02/r/  
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Fig. 14

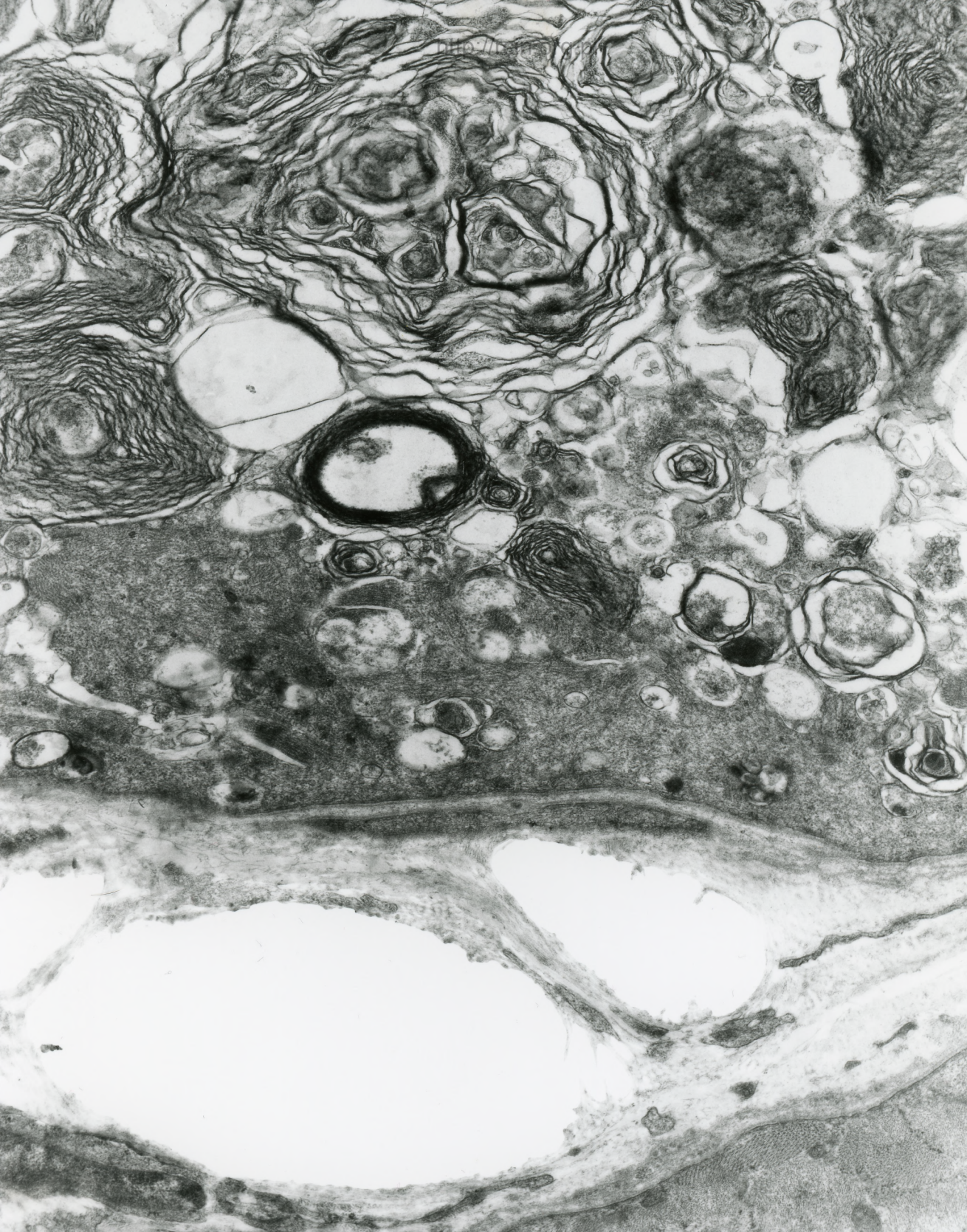




12/R  
AKU X6000

Fig.15

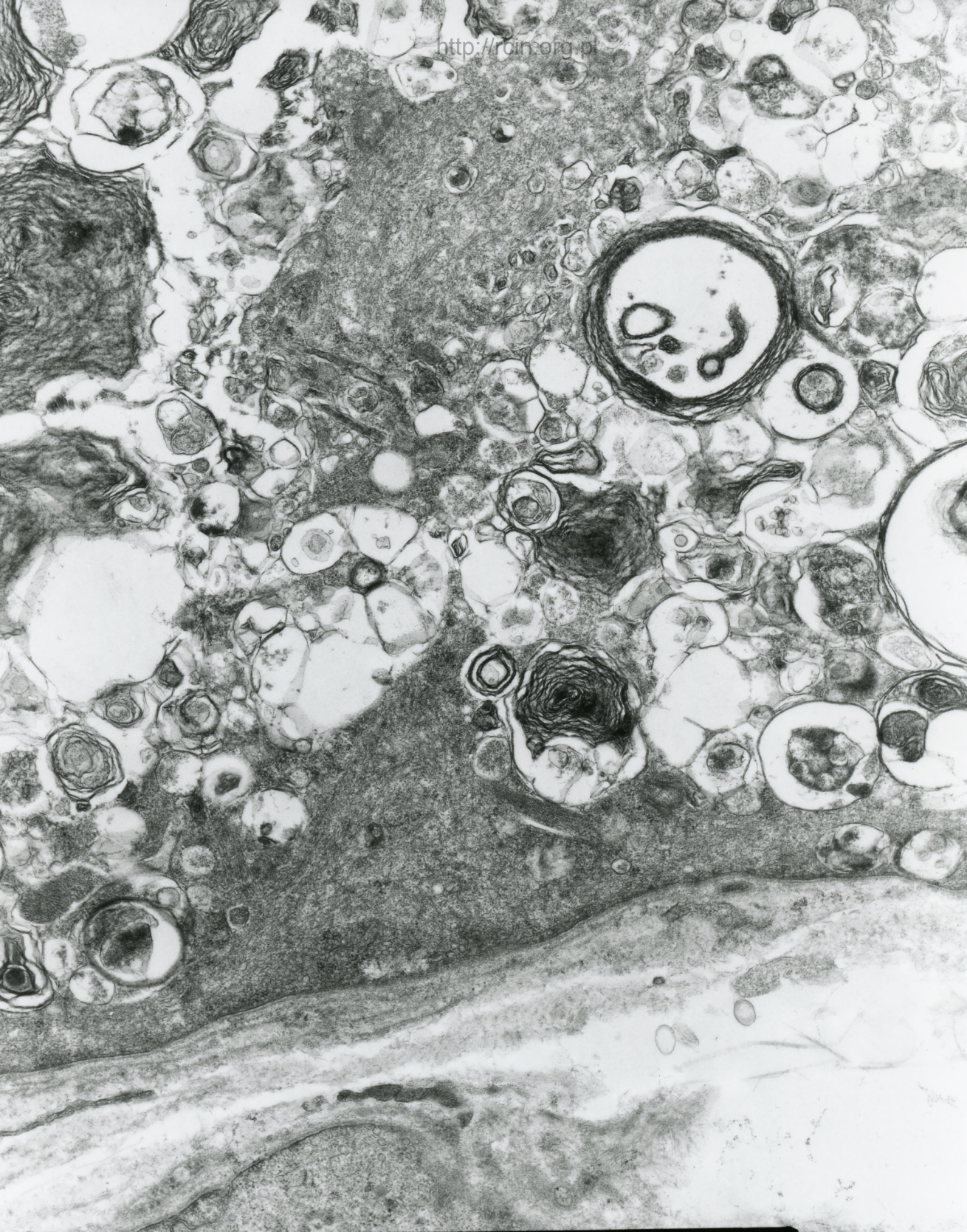




AF 10/02/1  
2R5459 80.0KV #X6000 1F#

Fig.16

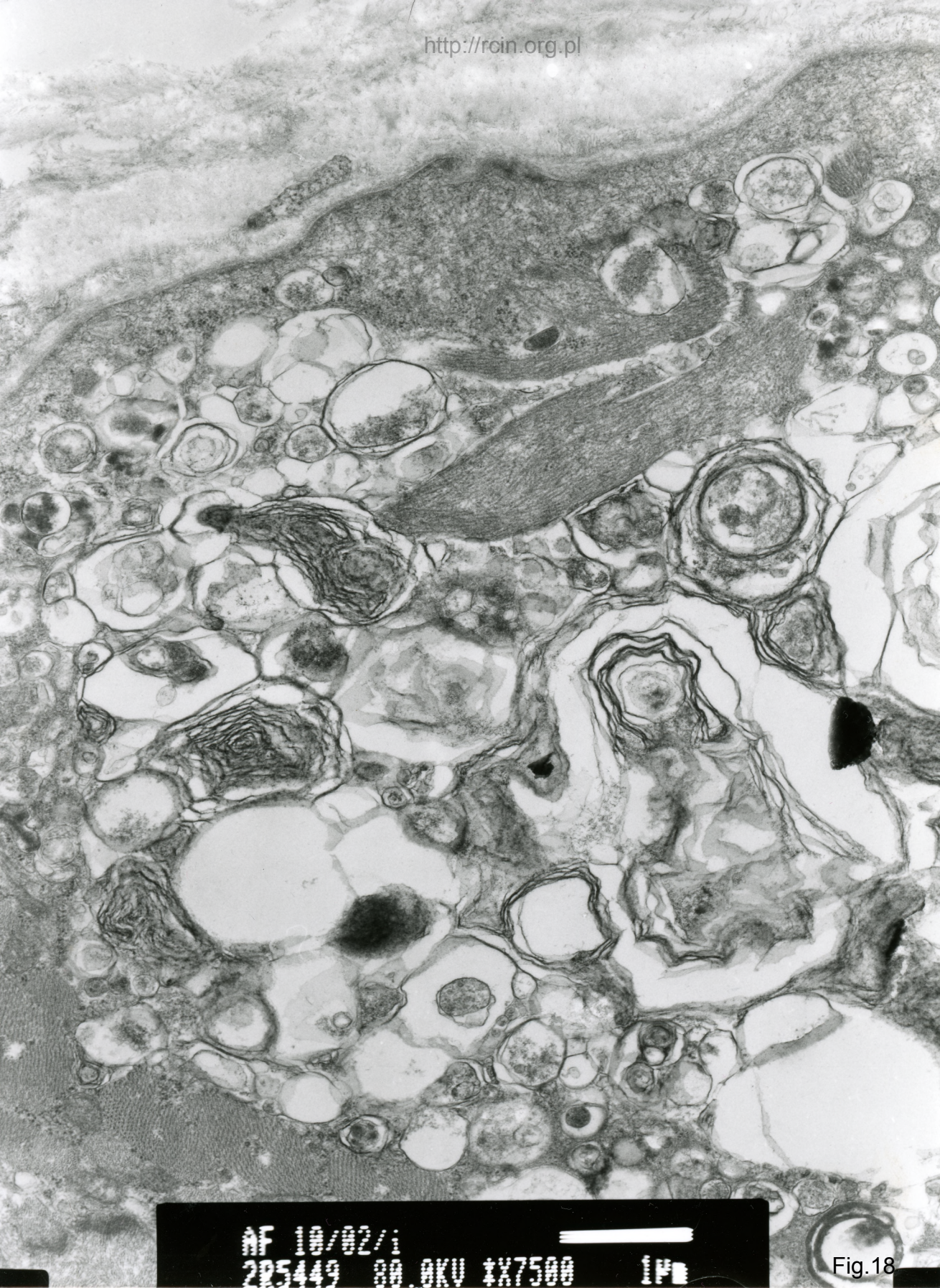




AF 10/02/i  
225460 00.0KV 1X6000 1P

Fig.17

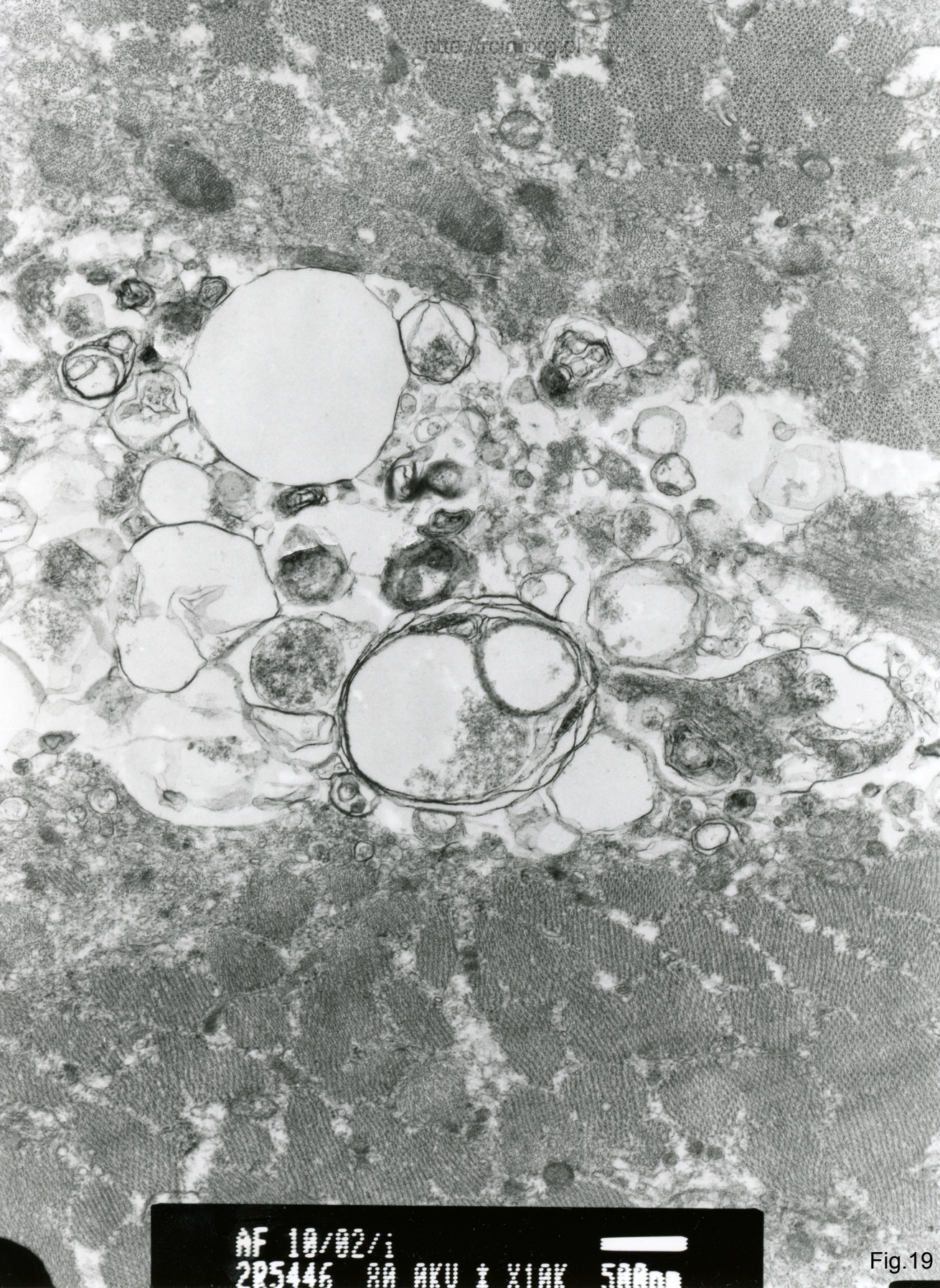




AF 10/02/1  
205449 80.0KV X7500 1P

Fig.18





AF 10/02/1  
285446 80 AKU ± XIAX 500nm

Fig.19

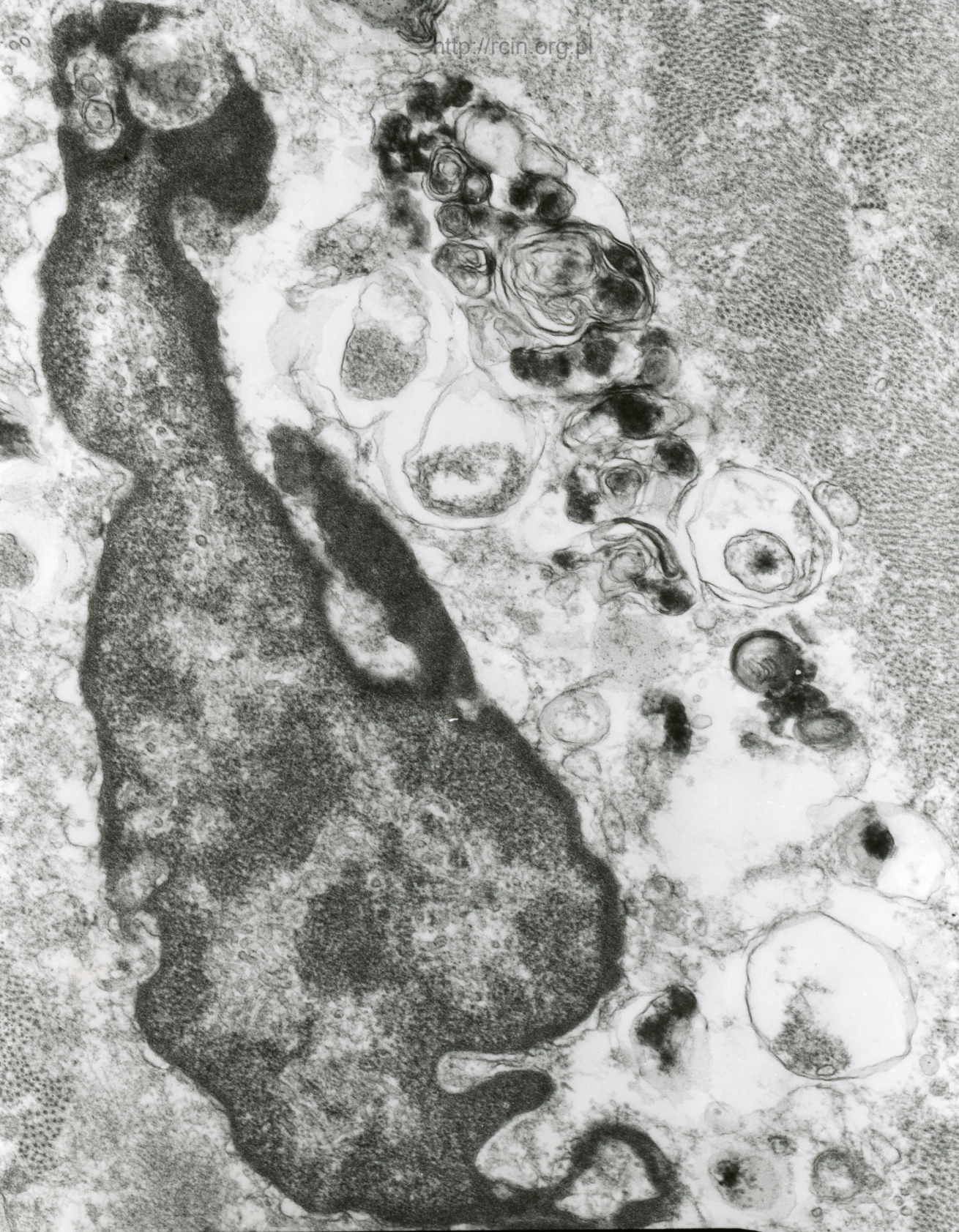




AF 10/02/0/  
2R5501 80.0KV ± X10K 500nm

Fig.20





HF 100/02/A  
05/11/7 80.0KV X15K 500nm

Fig.21

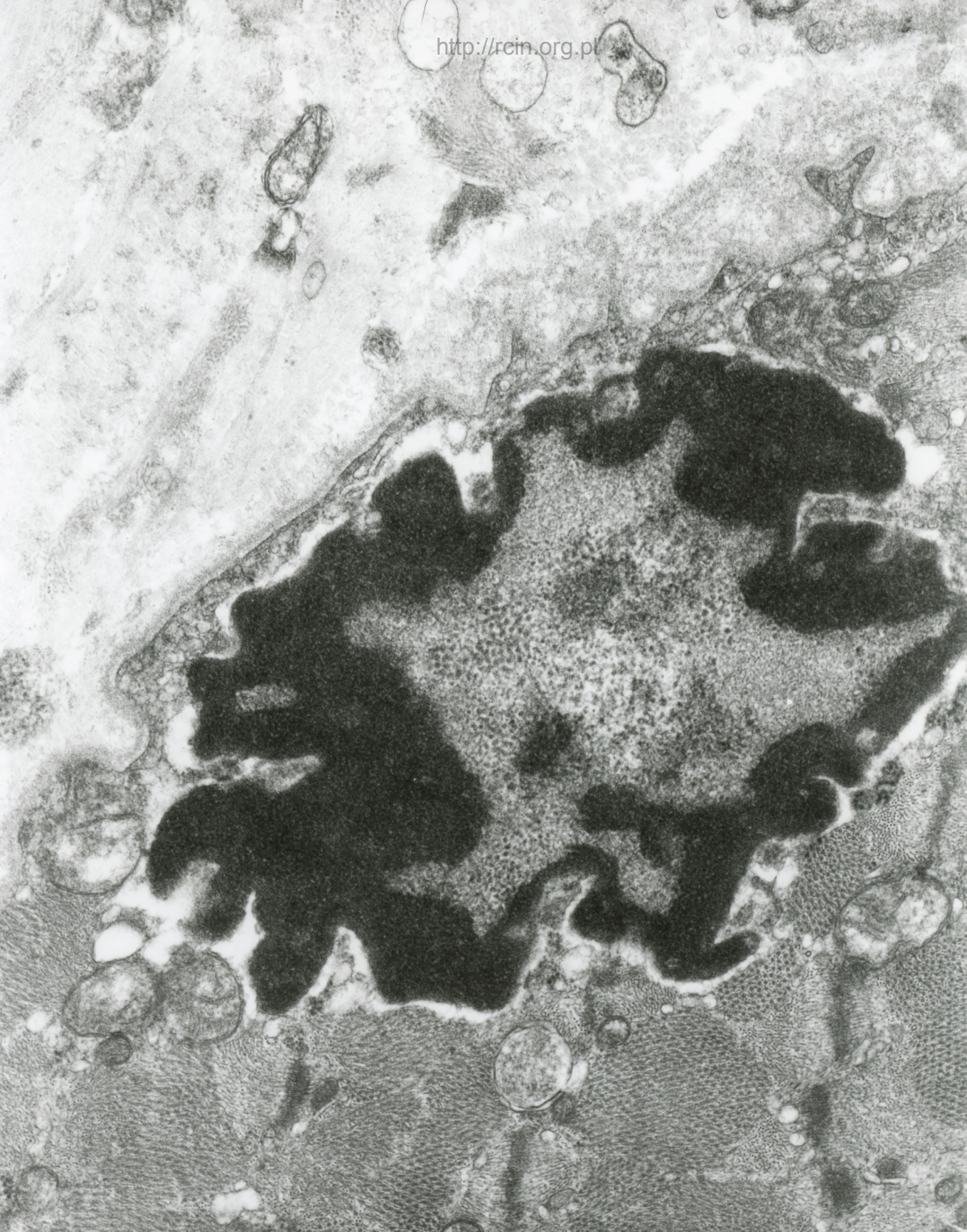




AF 19/02/0/  
205422 SA AKU 1X7500 1P

Fig.22





AF 10/02/W  
066060 80.0KV X15K 500nm

Fig.23





AF 10/02/1  
066022 80.0KV X15K 500nm

Fig.24

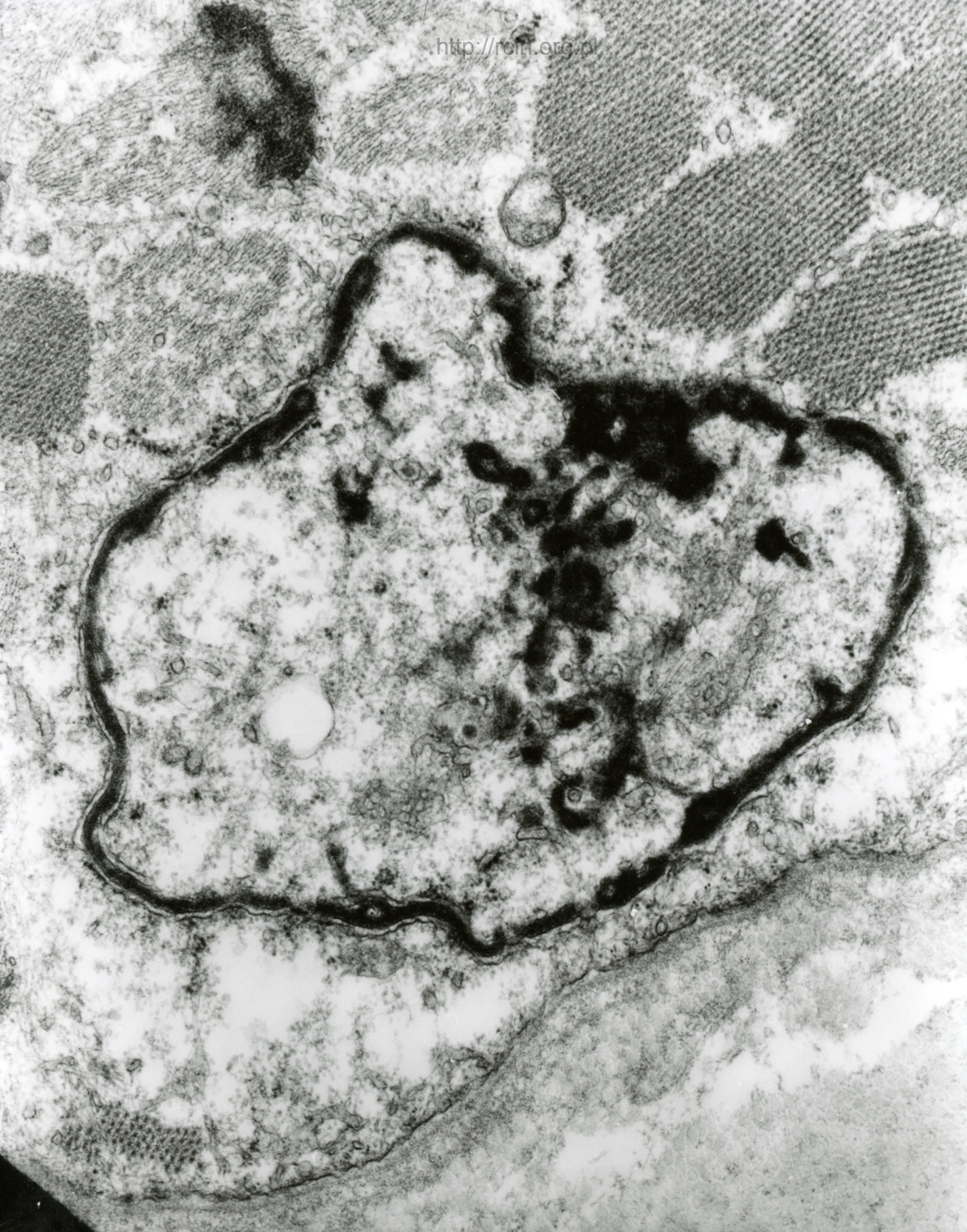




AF 10/02/R  
066020 80.0KV X15K 500nm

Fig.25





24 100000 02/R  
00.00V X15K 500nm

Fig.26





AF 110/02/R  
000000 00.0KV U20K 200um

Fig.27





AF 10/02/r/  
285593 80 AKU ± X12K 500nm

Fig.28

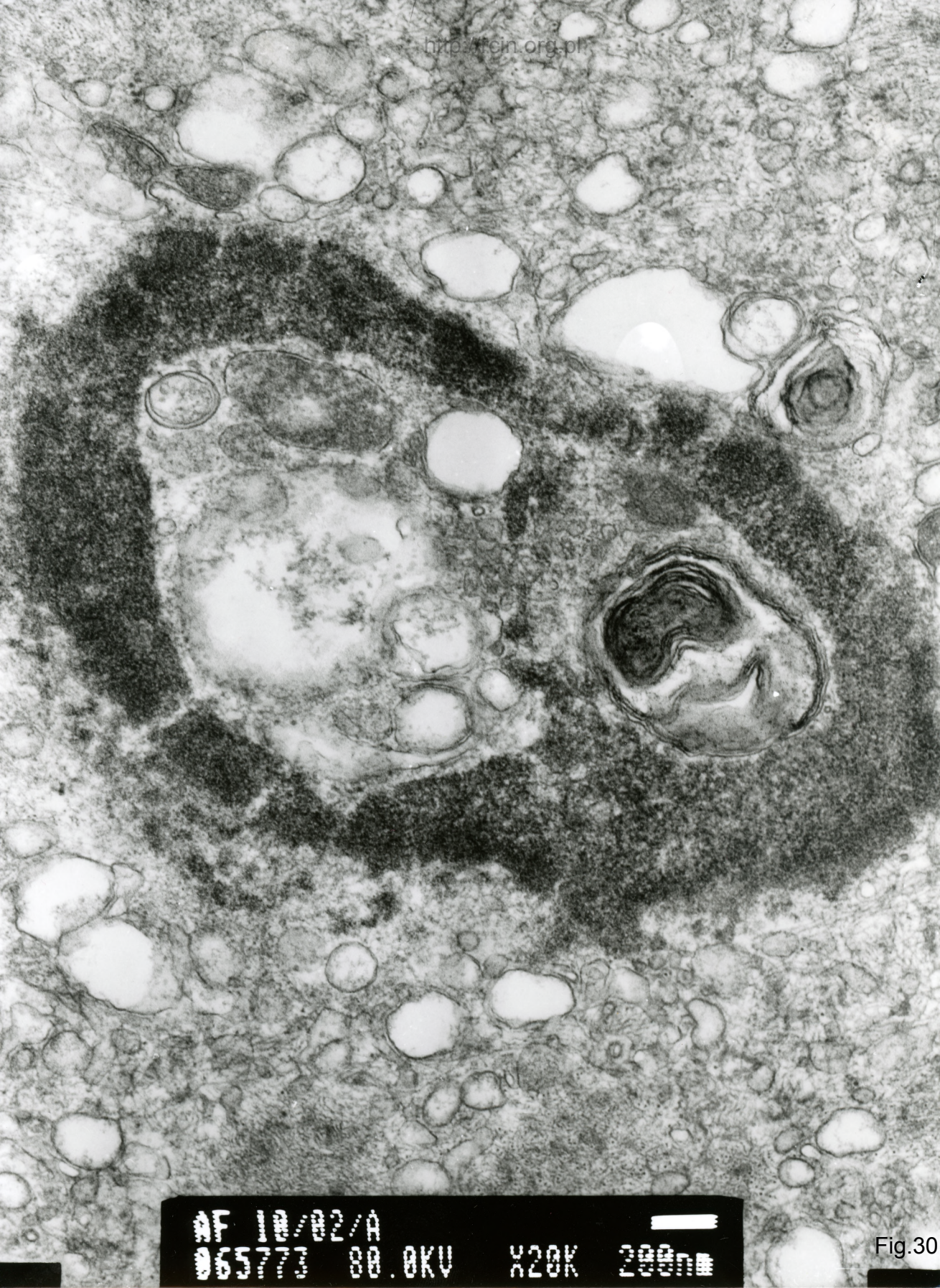




AF 10/02/0.  
225497 80.0KV X4000 ZF

Fig.29

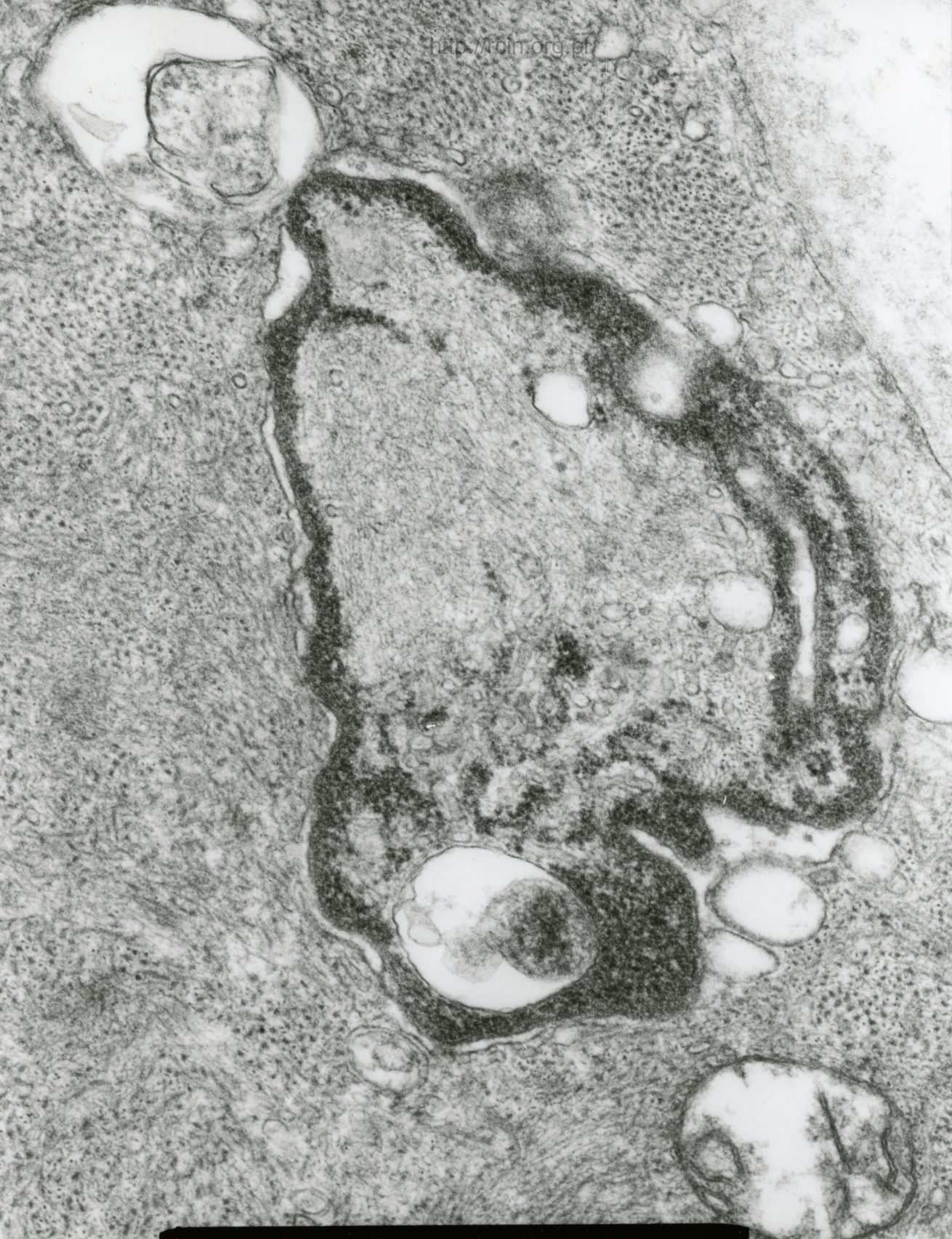




AF 10/02/A  
065773 80.0KV X20K 200nm

Fig.30





AF 10/02/J  
#55741 8A BKU U25K 200mm

Fig.31

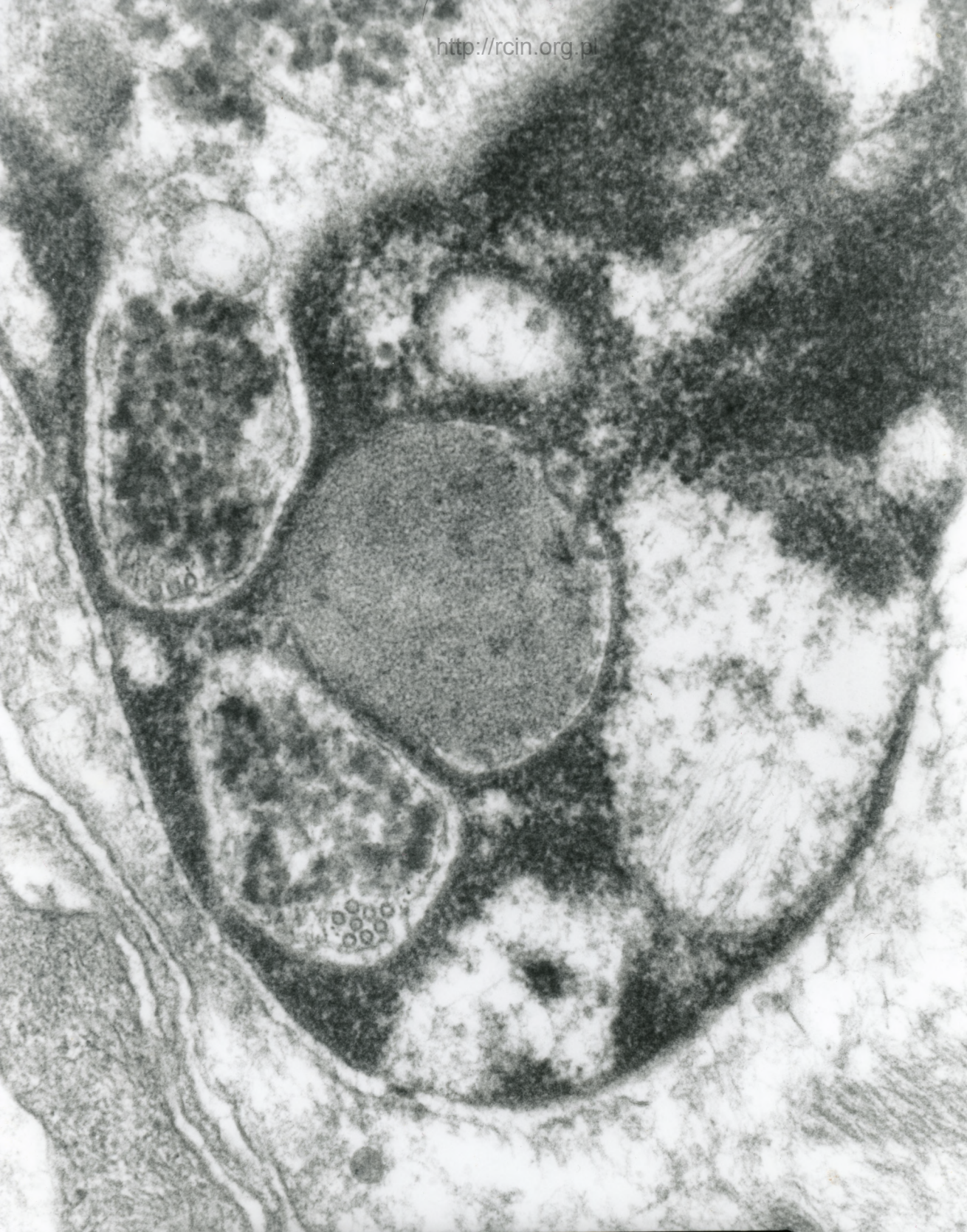




AF 10/02/J  
066053 80 0KV 015K 500nm

Fig.32

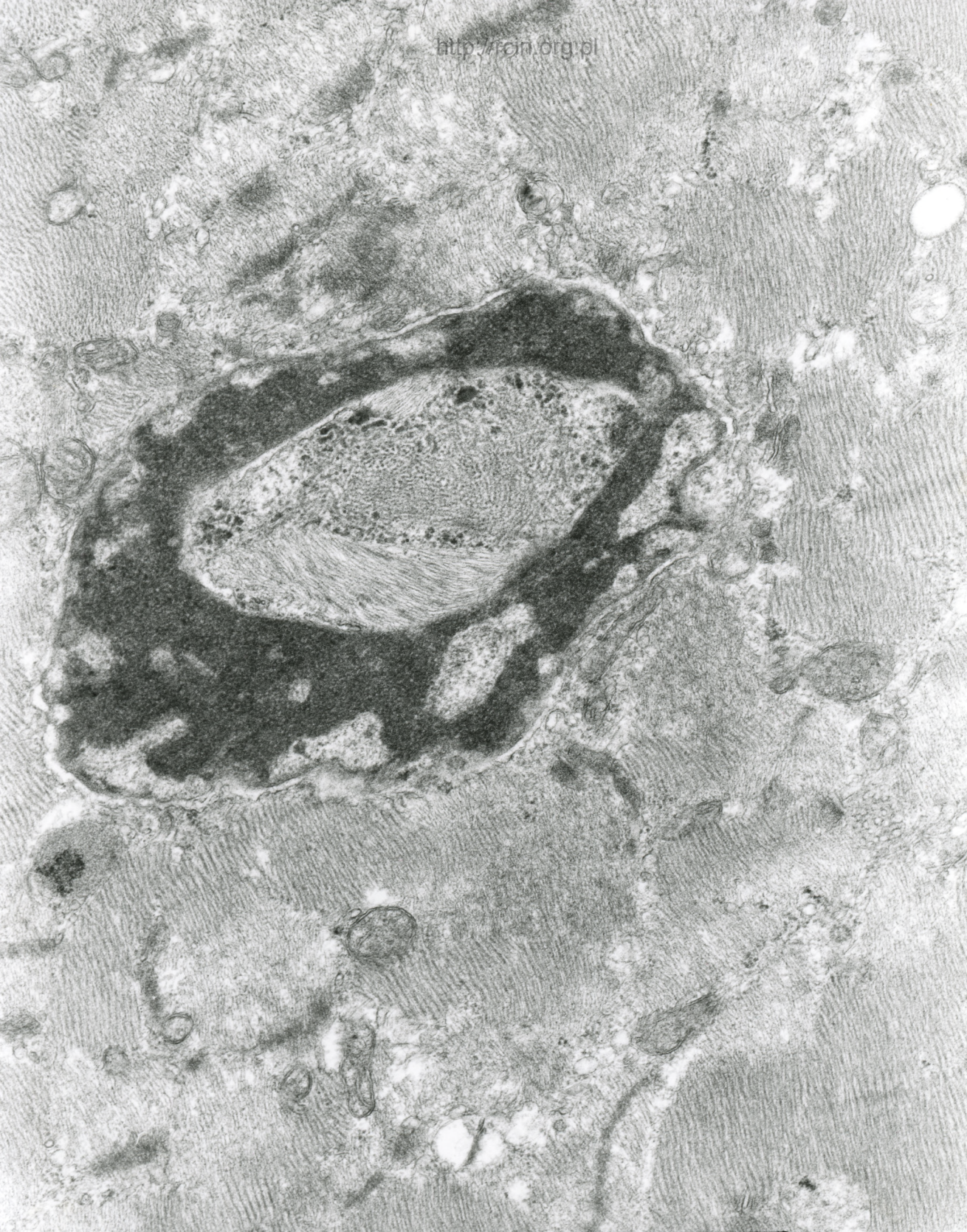




AF 10/02/1/  
2R5674 80.0KV X30K 200nm

Fig.33





AF 10/02/1  
205454 80.0KV X12K 500nm

Fig.34

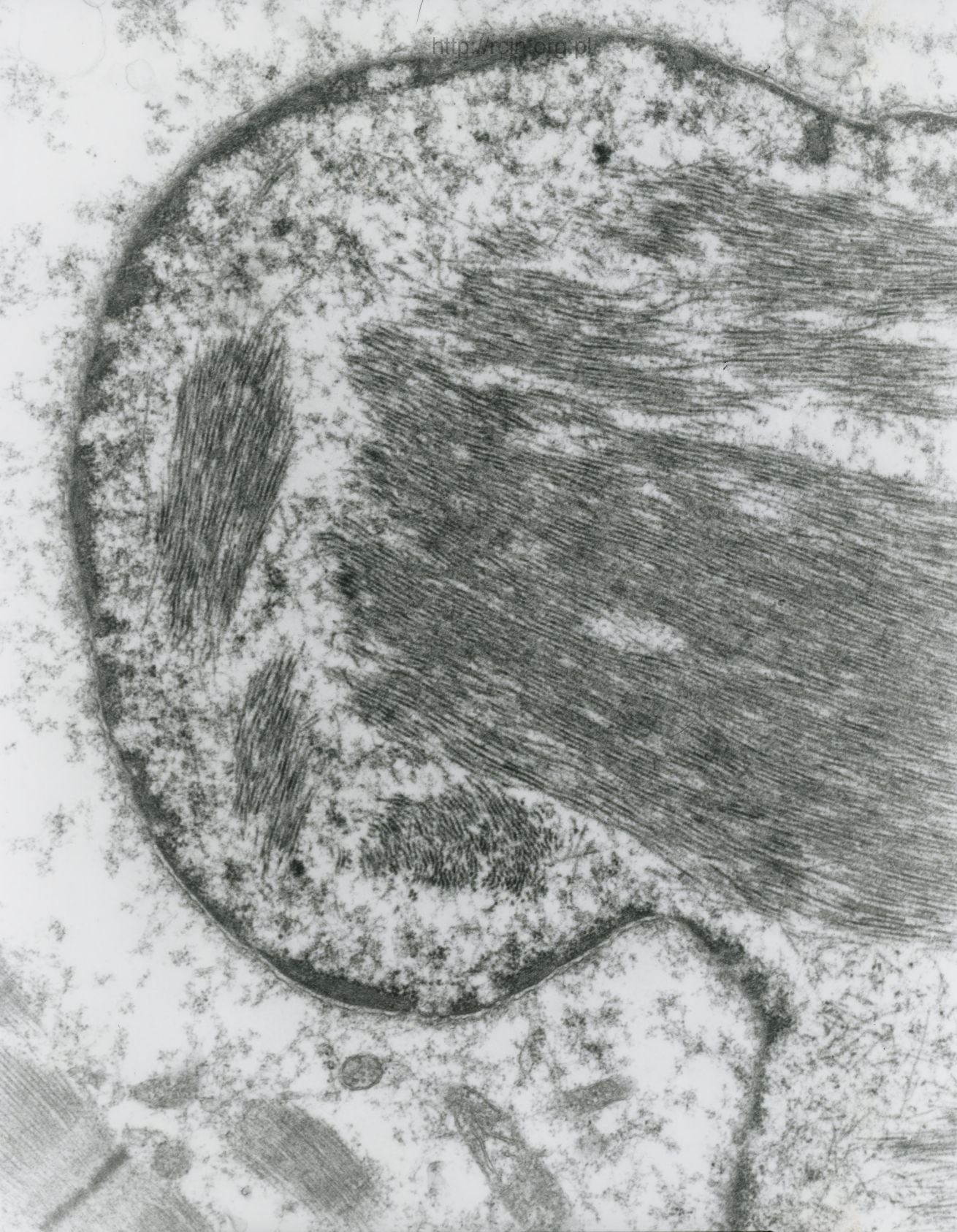




AF 10/02/v/  
2R5670 80.0KV ± X15K 500nm

Fig.35

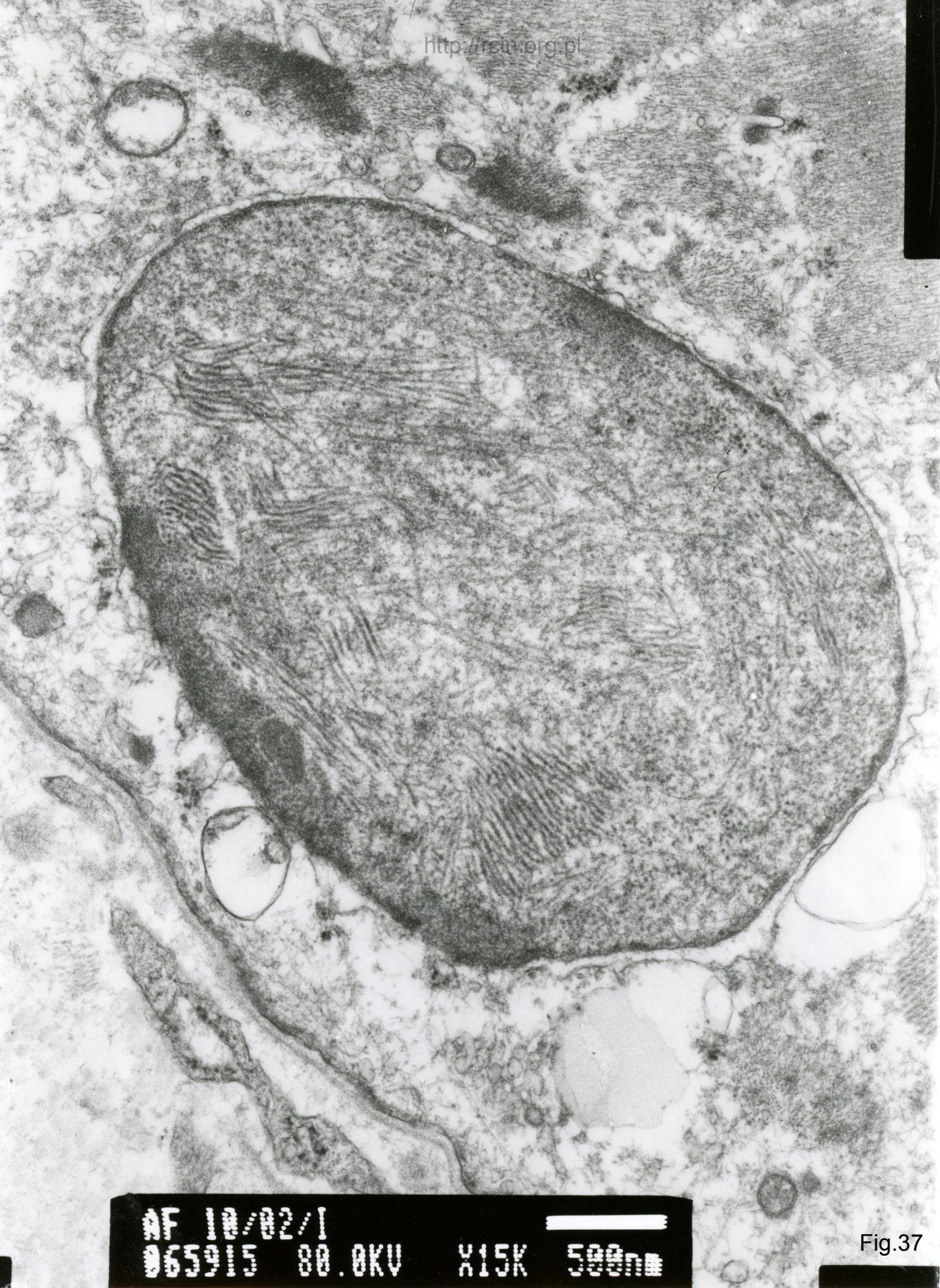




AF 10/02/r/  
205500 00 AKU + Y10K 500

Fig.36

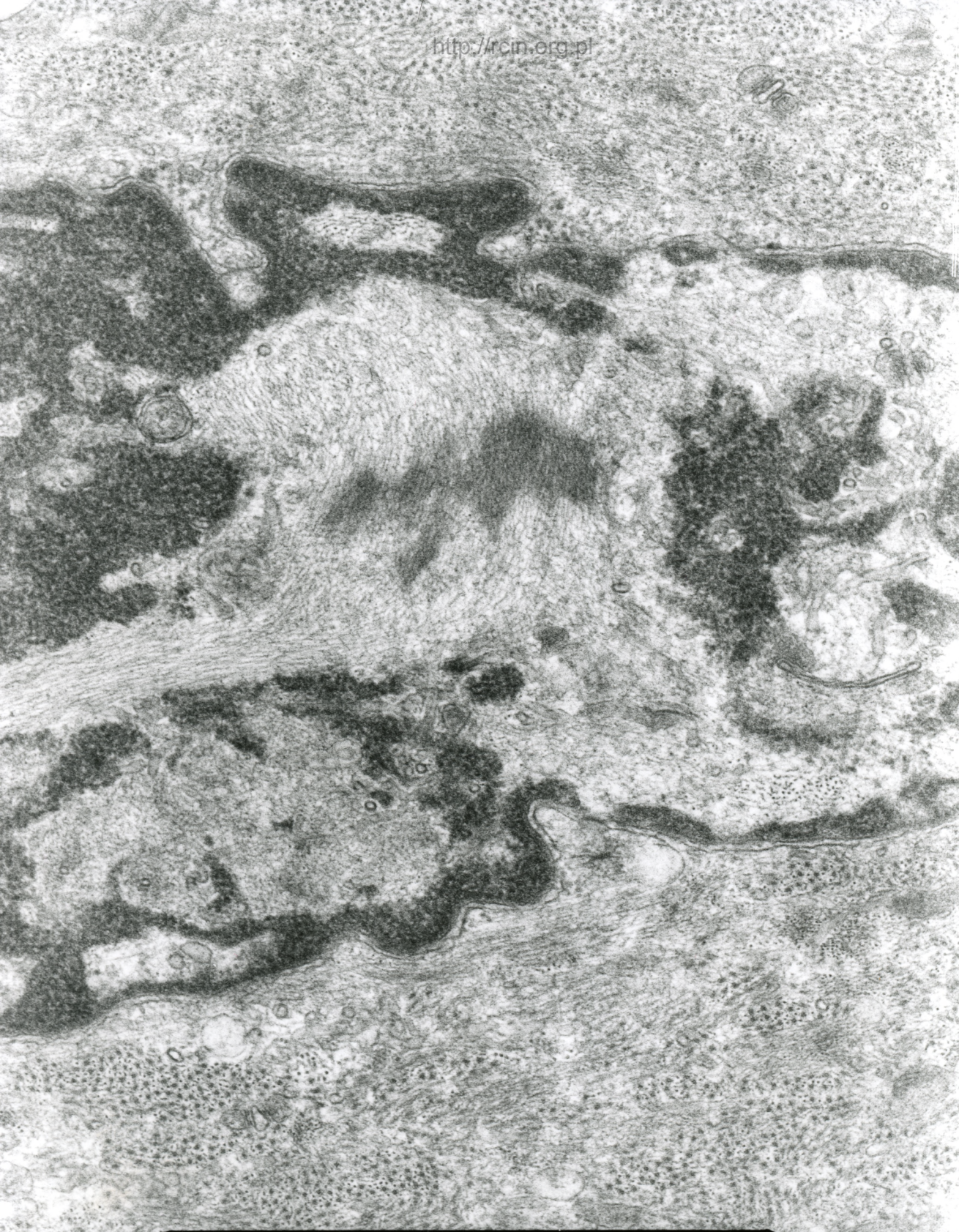




MF 10/02/1  
065015 80.0KV X15K 500nm

Fig.37

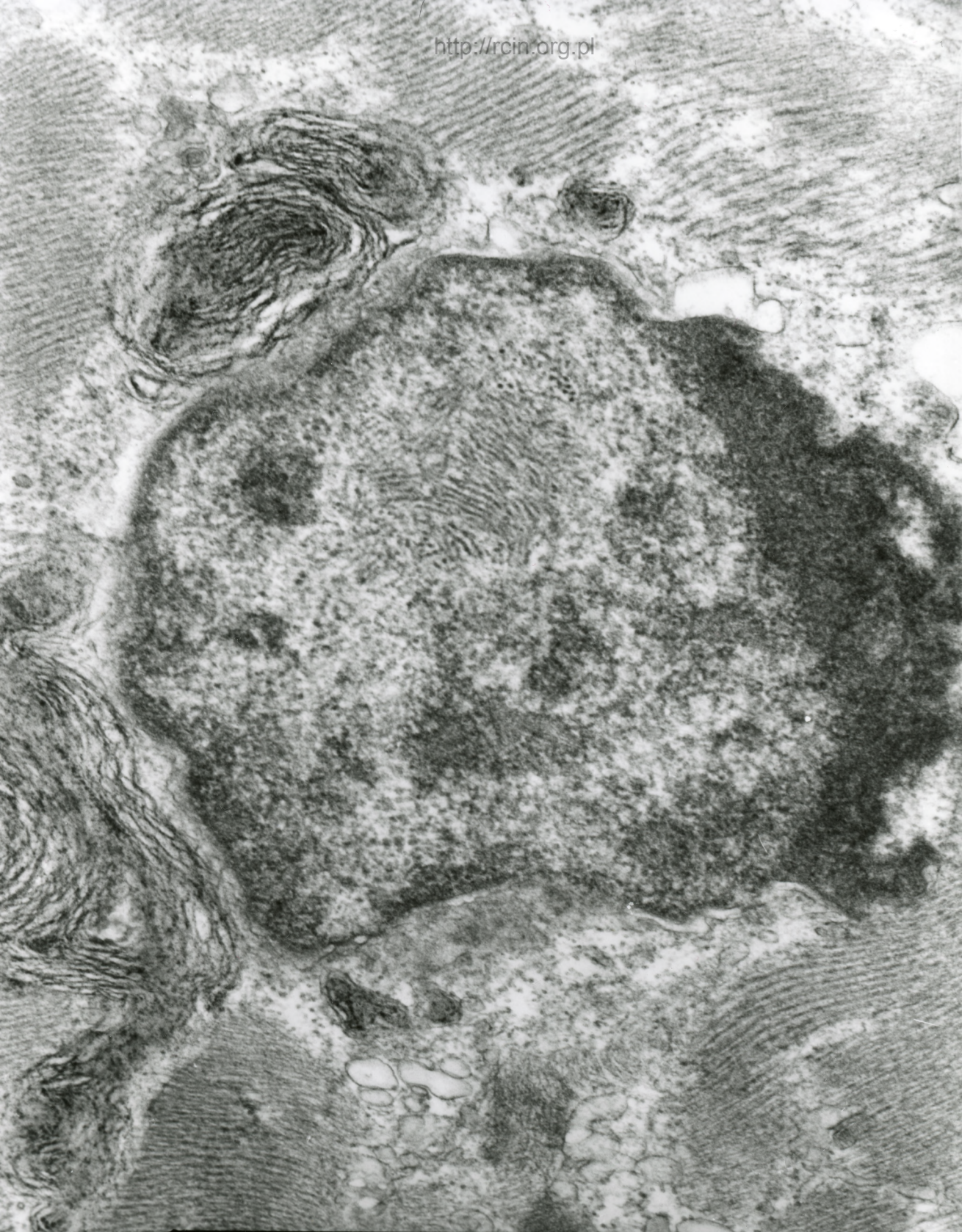




AF 10/02/z/  
205549 SA AKU ± X20K 200nm

Fig.38

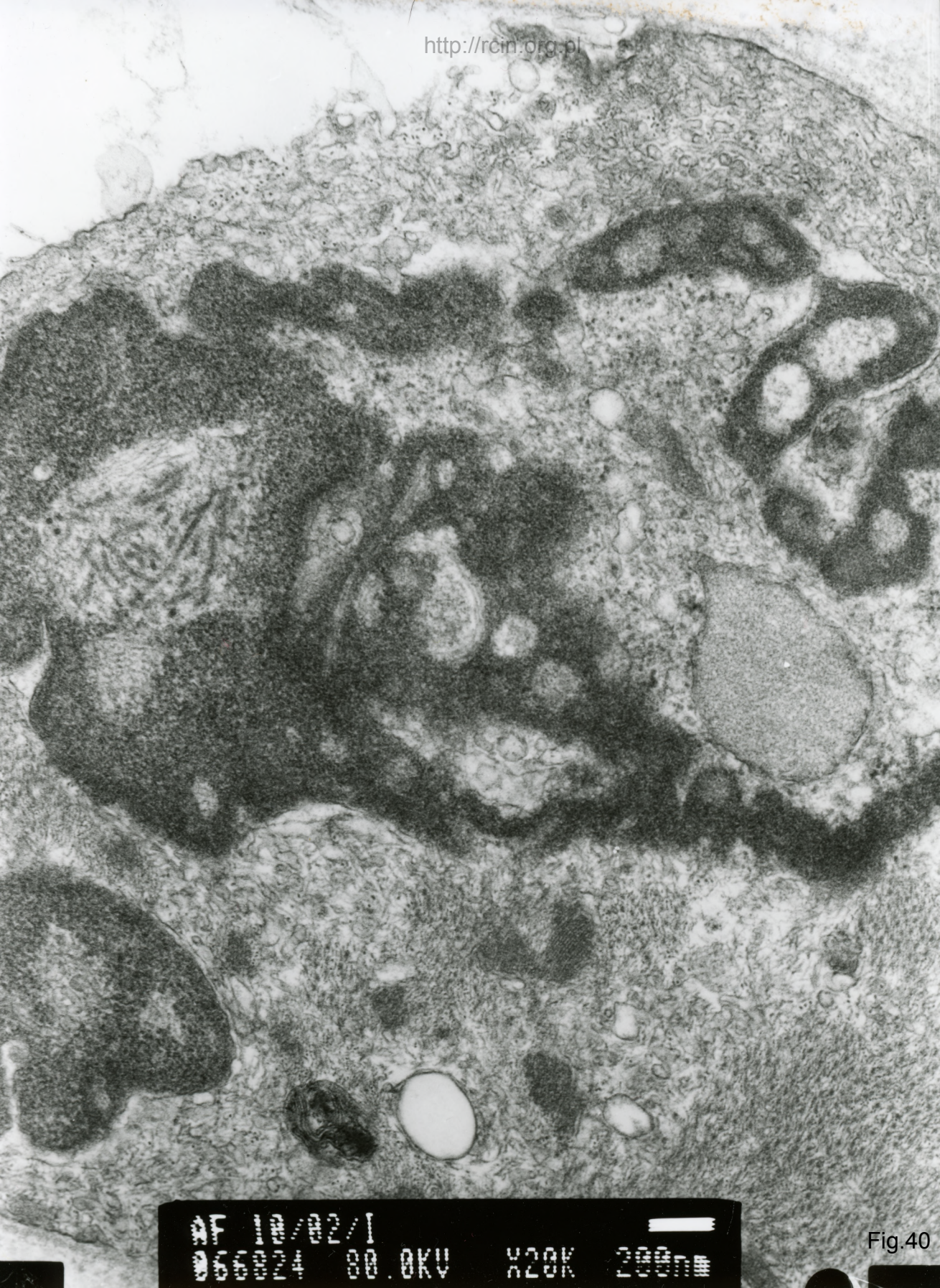




AF 10/02/1  
060020 80.0KV X20K 200nm

Fig.39

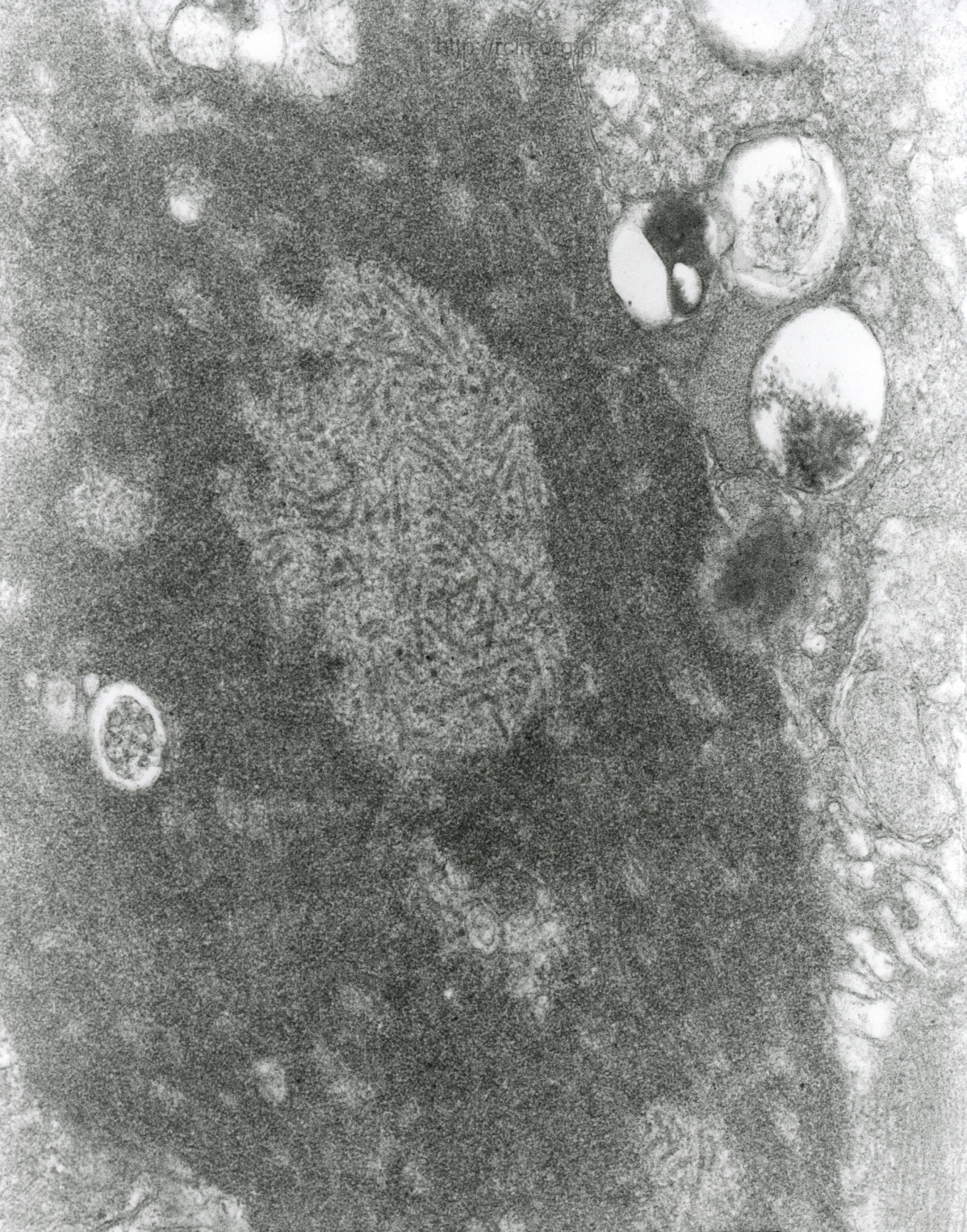




AF 10/02/1  
066024 80.0KV X20K 200nm

Fig.40

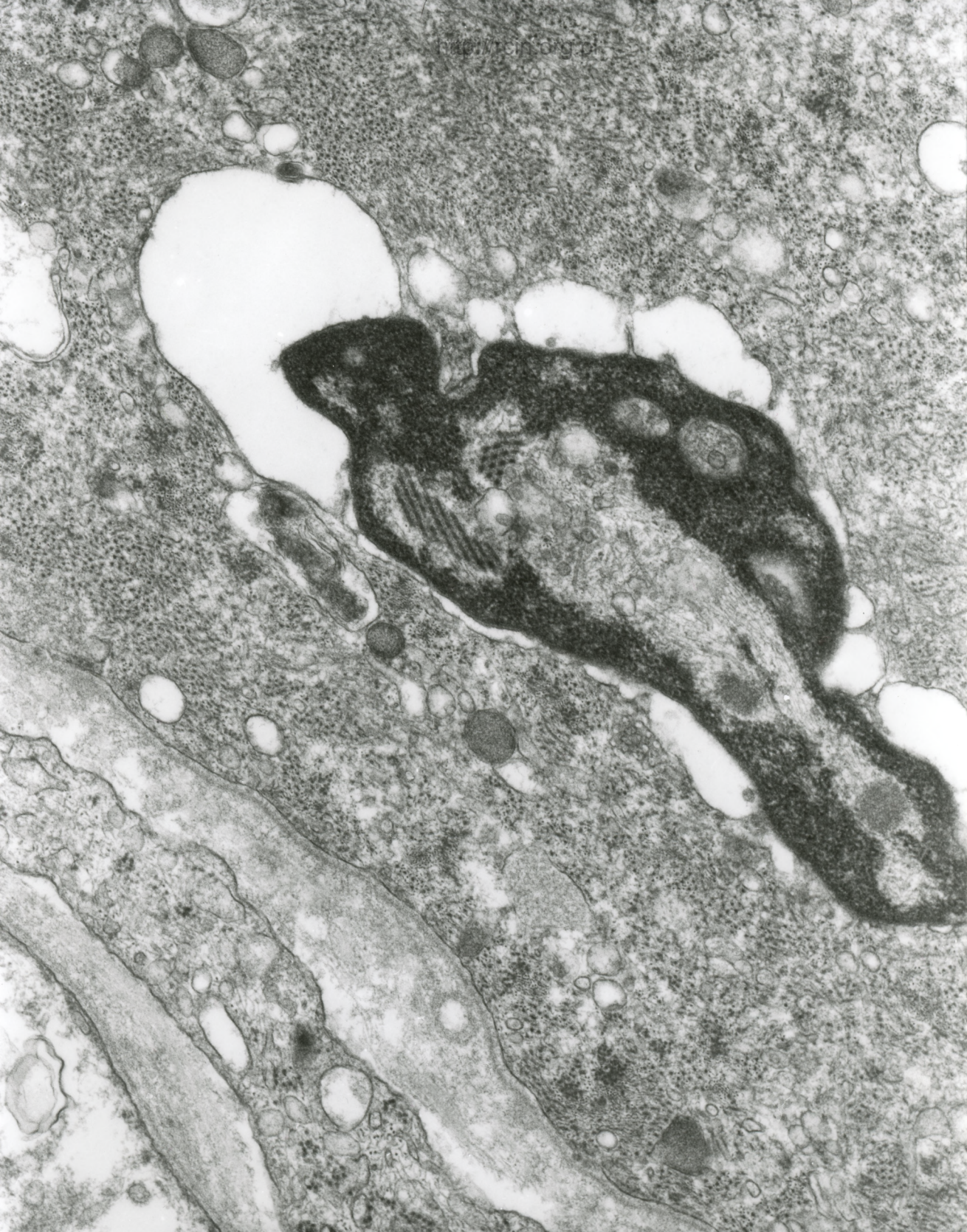




AF 10/02/0/  
205499 00.0KV ± X20K 200nm

Fig.41

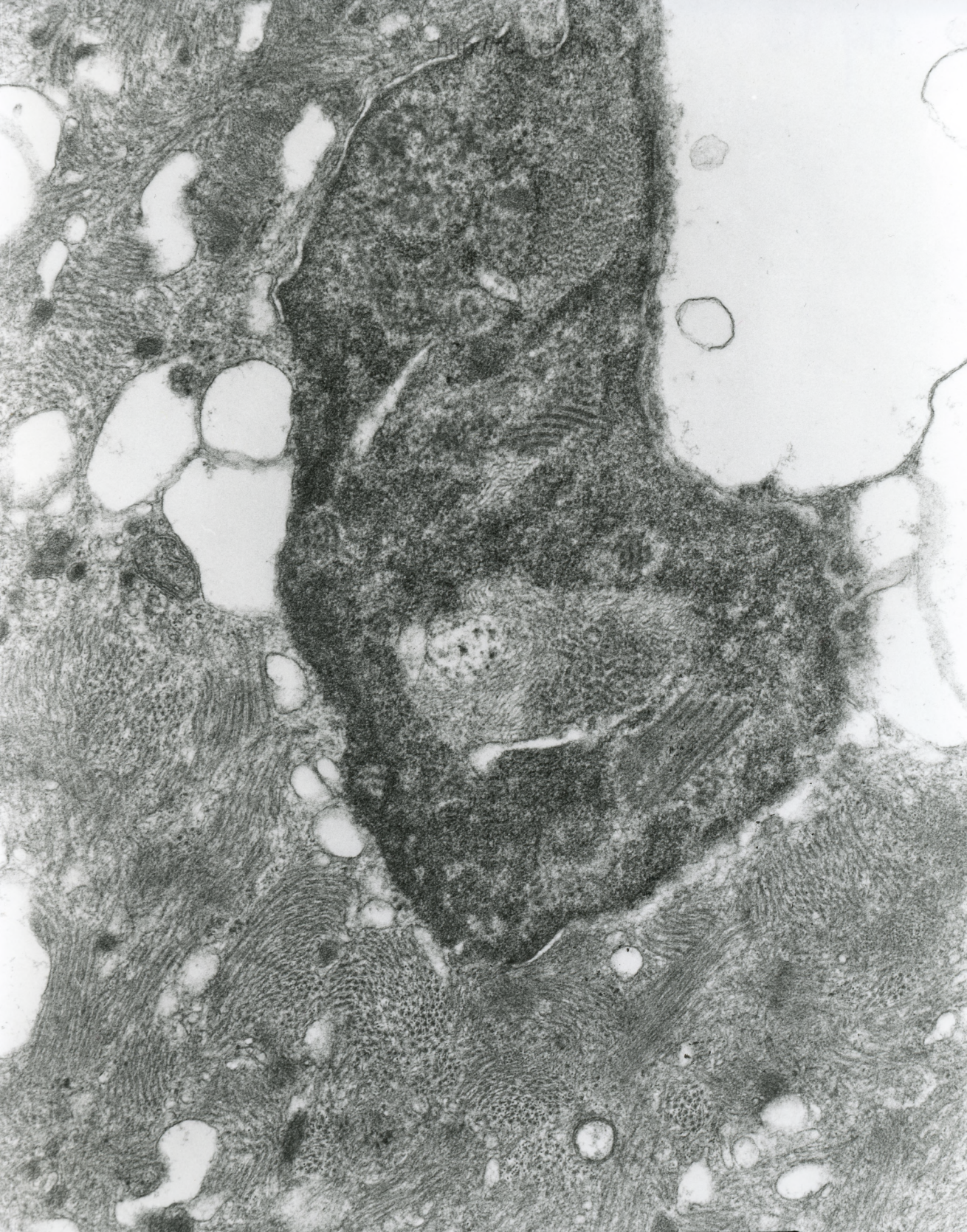




AF 10/02/J  
066052 80.0KV X15K 500nm

Fig.42

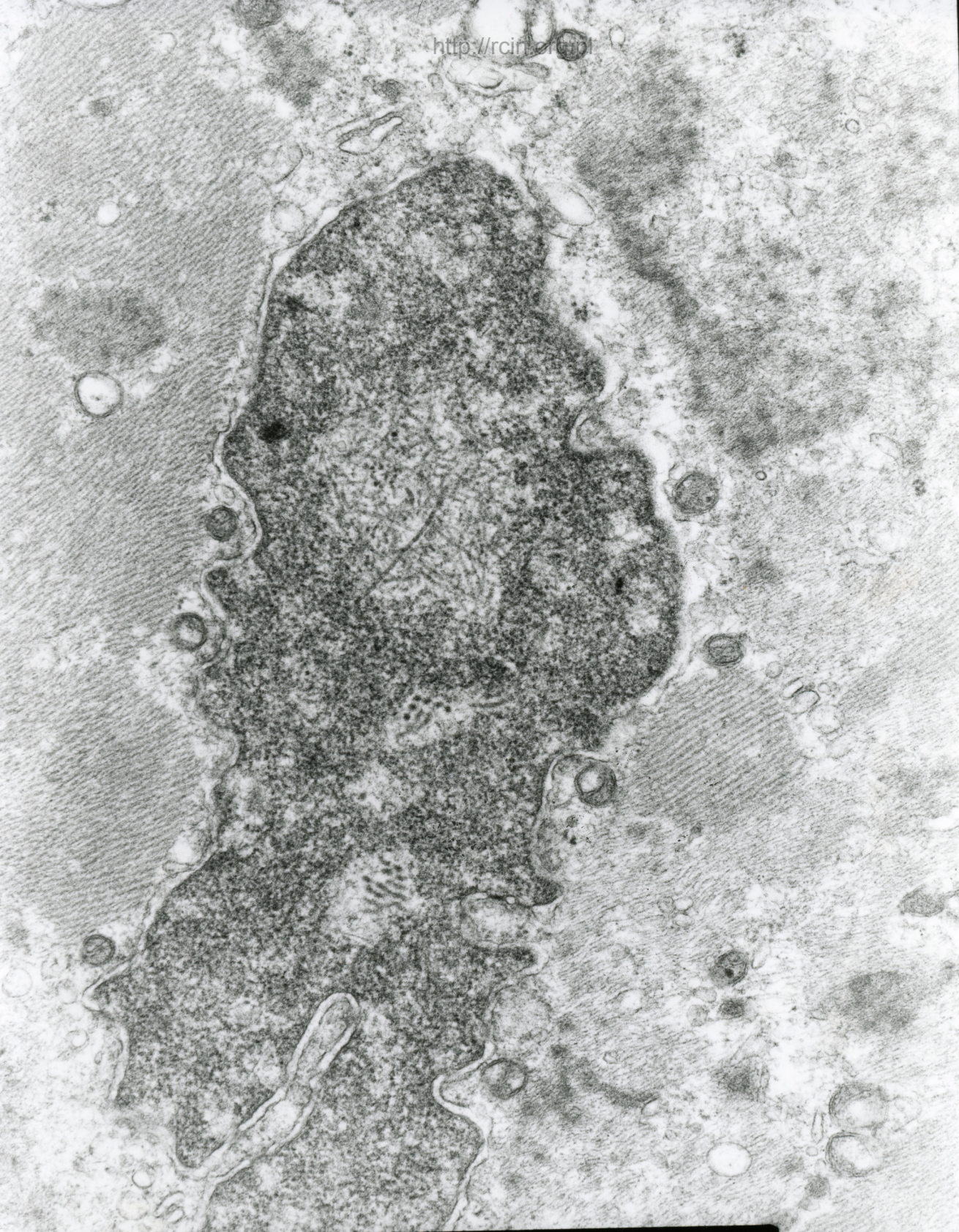




AF 10/02/T/  
225485 80.0KV † X15K 500nm

Fig.43

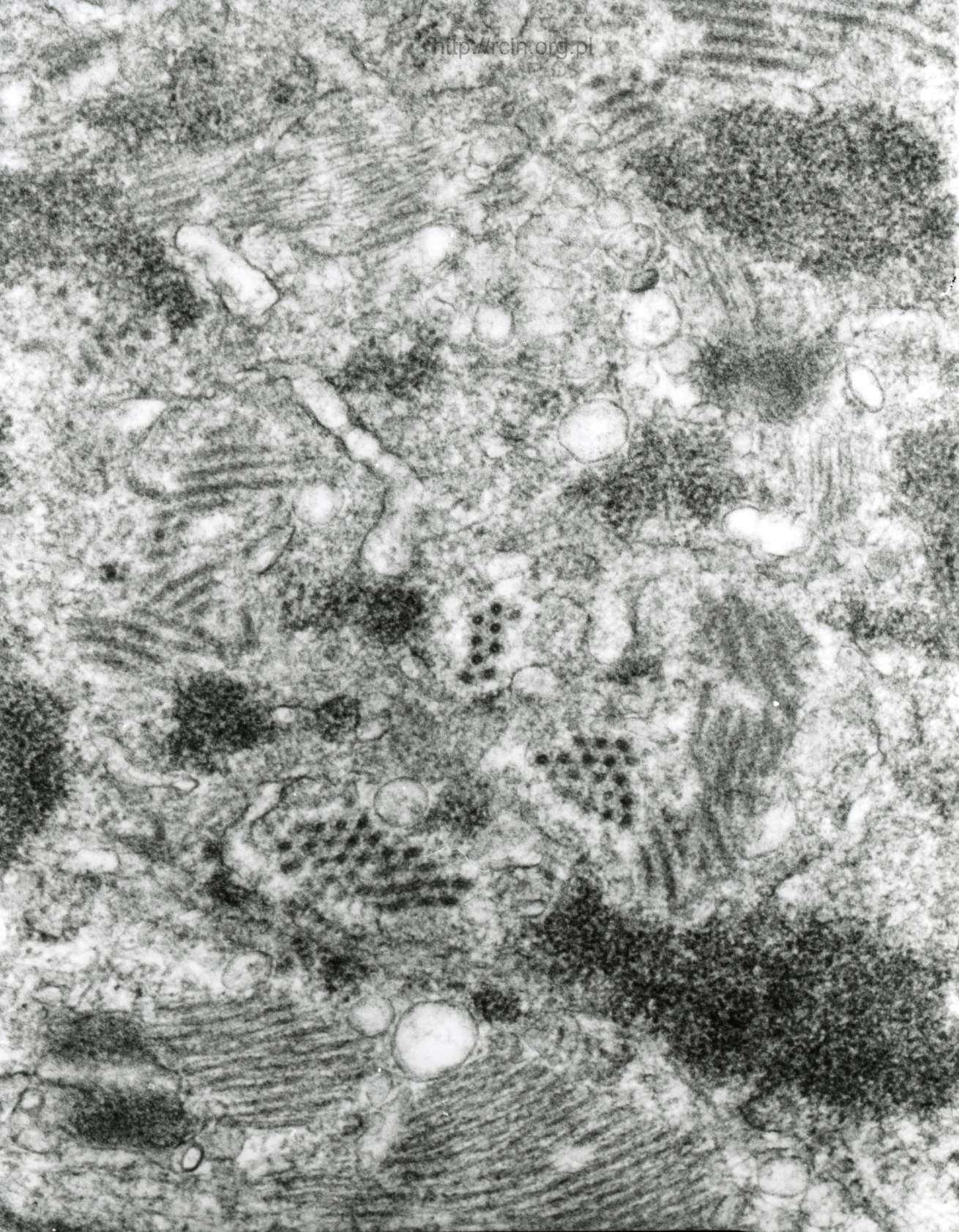




AF 10/02/i/  
2P5657 8A AKU ± X15K 500nm

Fig.44

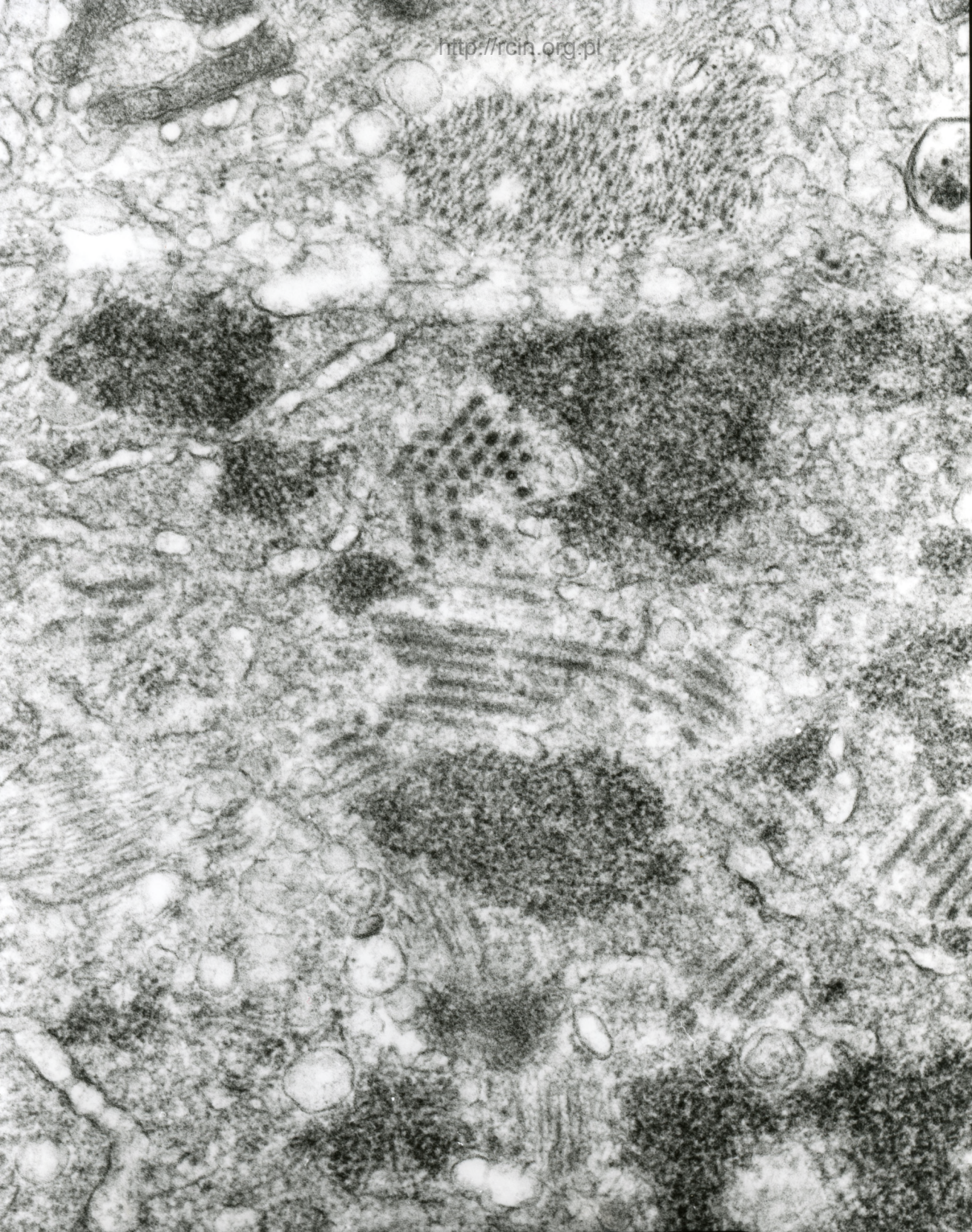




AF 10/02/1  
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Fig.45

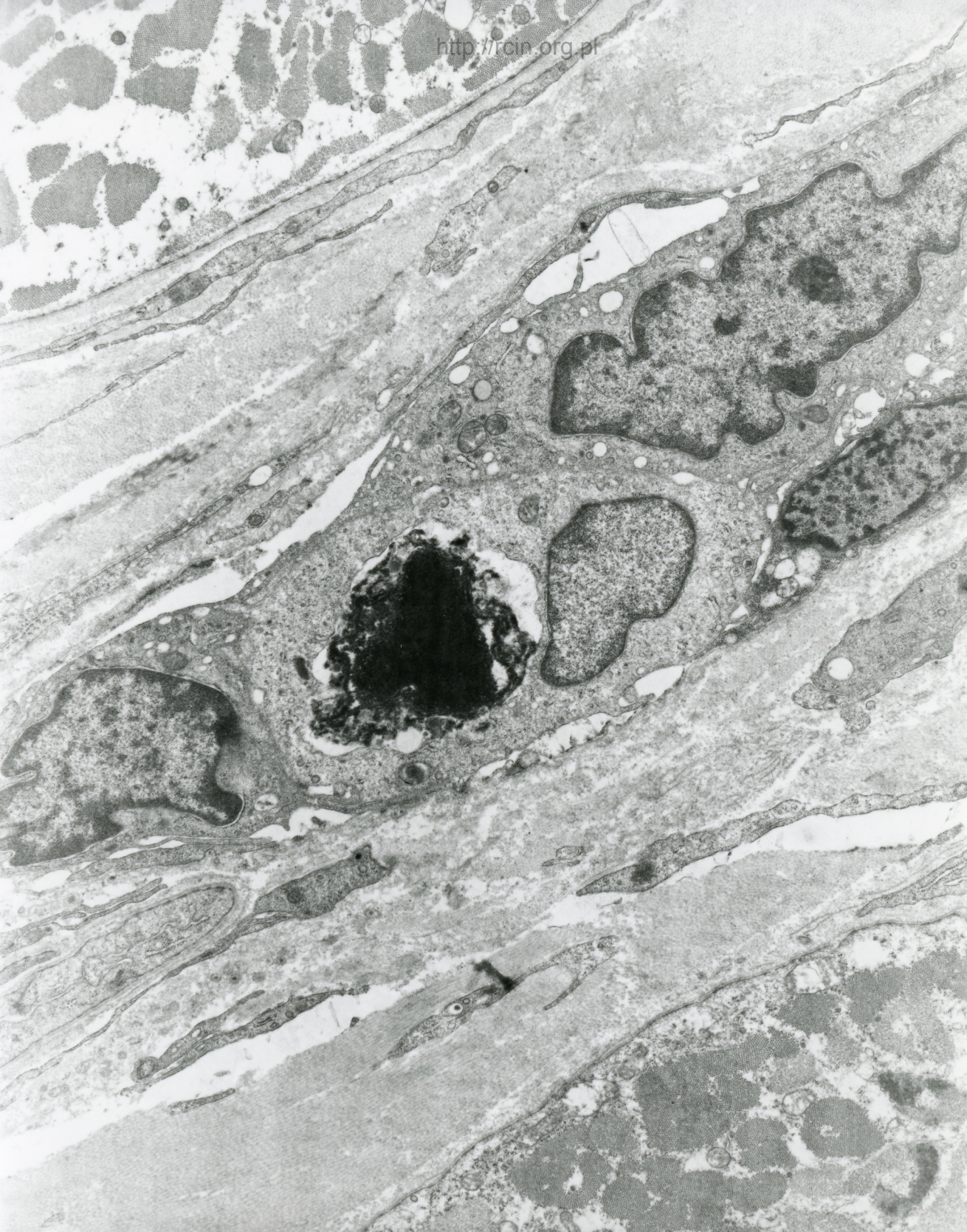




AF 10/02/1  
065767 80.0KV X30K 200nm

Fig.46





AF 10/02/1/  
2R5655 00.0KV 1X3000 2P0

Fig.47





AF 10/02/r/  
205509 30.0KV 1X3000 2P

Fig.48



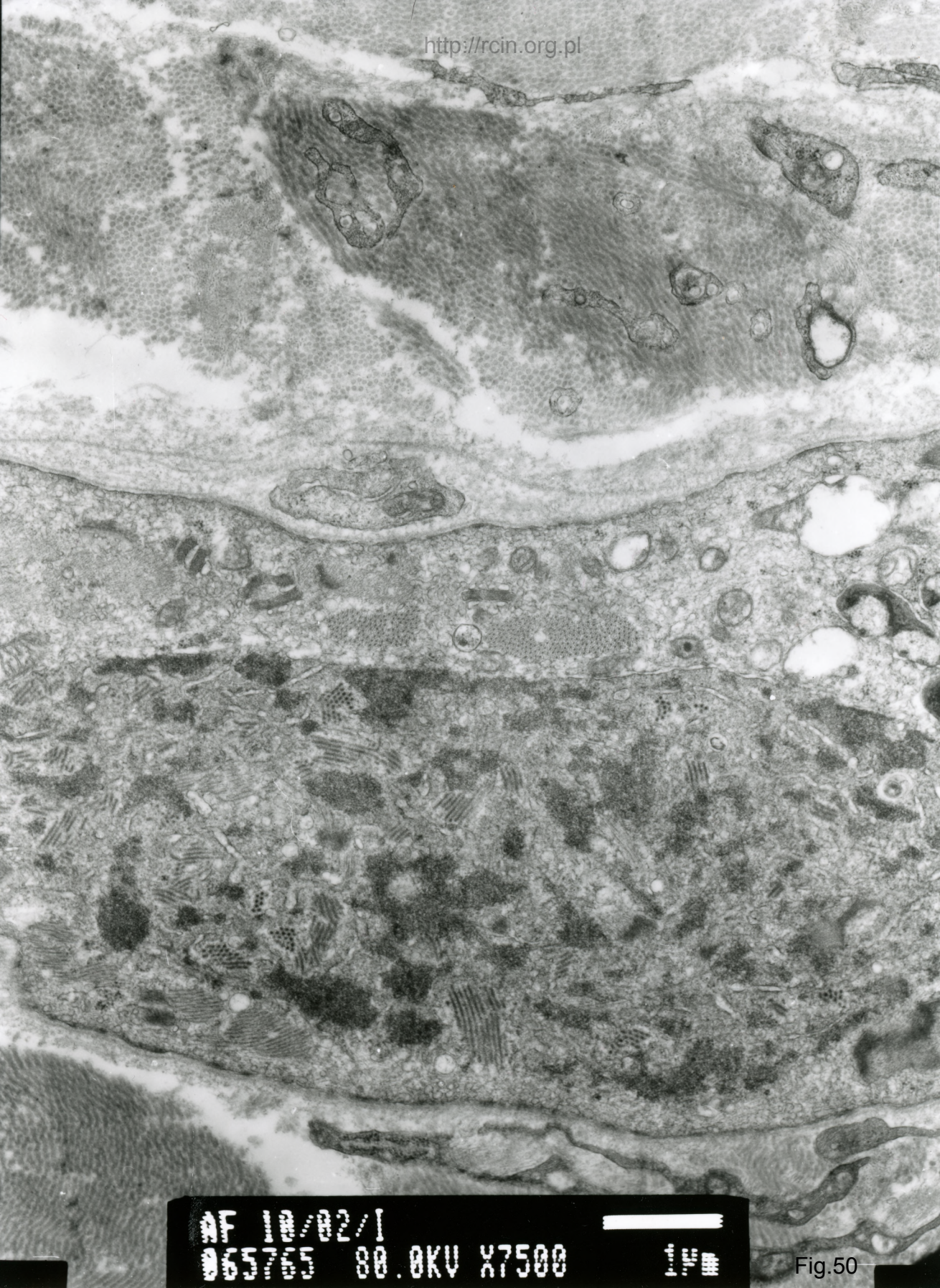


AF 10/02/r/  
2R5592 80.0KV  $\times$ 5000

1P

Fig.49





AF 10/02/1  
065765 80.0KV X7500

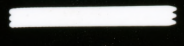


Fig.50