Microvascular pathology of Friedreich cardiomyopathy

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Nikolaus Friedreich (1877) was aware of heart disease in his patients but thought it was unrelated to the neurological disorder. In 1946, Dorothy Russell considered cardiomyopathy an integral part of Friedreich ataxia (FA). In addition to sparse inflammatory infiltration, sections show fibrosis and capillary hyperplasia. We examined the left ventricular walls of 41 homozygous FA patients aged 10–87 and 21 controls aged 2–69. An antibody to CD34 enabled quantitative capillary profile counts for a comparison with cardiomyocyte counts in the same field. Mean capillary counts in normals were 1926±341/mm², and the median ratio of capillaries to cardiomyocytes was 1.0 (interquartile range [IQR]: 0.9–1.2). In FA, however, the number of cardiomyocytes/mm² was less, and the median ratio of capillaries to heart fibers was 2.0 (IQR 1.4–2.4). There was a significant correlation of the higher guanine-adenine adenine-trinucleotide length (shorter allele, GAA1) with the younger age of onset, shorter disease duration, and lower cardiomyocyte counts. The ratio of capillaries to heart fibers was higher in patients with long GAA1 repeat expansions (e.g., 3.31 in GAA1 of 1200). Double-label immunofluorescence for CD34 and S100A4 revealed co-expression in endothelial cells, supporting endothelial-to-mesenchymal transition in the pathogenesis of cardiac fibrosis (supported by Friedreich’s Ataxia Research Alliance).

LEARNING OBJECTIVES

The presentation will enable the learner to:

1. Describe endothelial-to-mesenchymal transition in the pathogenesis of cardiac fibrosis in Friedreich cardiomyopathy

Non-Perfused Brain and Retino-Dural Hemorrhage

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10 cases of pneumonia causing cardiac arrest and non-perfused brain occurred at ages 40 days-30 months, in a medico-legal setting. In each deceased child, both the pneumonia and non-

LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Explore the current theories leading to neonatal death in prolonged labor
2. Summarize the known pathological findings associated with vacuum and forceps
3. Discuss the significance of intraparenchymal hematoma in the setting of prolonged delivery

Parasagittal intraparenchymal hemorrhage in complicated second stage labour: a report of three cases

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The increased use and mastery of cesarian section for deliveries and the refinement of technologies for assisted delivery in the setting of difficult second stage of labour have made intrapartum deaths more rare and modern obstetrical pracices are rarely accompanied by the classic forceps related intracranial injuries. We document a novel pattern of intracranial injury in three cases of neonatal death following prolonged labor, of which two out of three required vacuum and forceps.

All three showed similar bilateral parasagittal intraparenchymal haemorrhages and cerebral edema, in a pattern reminiscent of “gliding contusion, as well as subgaleal haemorrhage of varying amout. Two out of the three cases showed parietal bone fractures and one demonstrated extensive calvarialuence. We briefly discuss the significance of these findings and implications for future cases.

LEARNING OBJECTIVES

This presentation will enable the learner to:

1. Discuss how PET-PiB beta-amyloid imaging is used as a potential biomarker of Alzheimer disease (AD)
2. Correlate postmortem neuropathologic evidence of beta-amyloidosis with PET-PiB data, and learn that PET-PiB is a potentially useful tool to detect beta-amyloidosis in presymptomatic and symptomatic individuals