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Nesidioblastosis: Ultrastructural and Immunohistochemical Observations

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Although hypoglycemia is a frequent occurrence in the newborn, persistent hypoglycemia due to hyperinsulinemia is rare. An early diagnosis goes a long way in

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preventing brain damage and subsequent mental retardation. A case of nesidioblastosis with complete electronmicroscopy and immunohistochemistry is reported in this communication. The hypoglycemia was resistant to diazoxide but responded to subtotal pancreatectomy. This is, to the best of our knowledge, the third reported case from the Indian subcontinent(1,2).

Case Report

A 5-day-old term LGA male Muslim newborn of nonconsanguineous parents presented with generalized tonic and clonic seizures. The birth weight was 4000g and the vaginal delivery prolonged. A previous term LGA sibling had expired on day 15 of an unestablished cause. Random blood sugar levels were persistently low and a continuous infusion of 12.5% dextrose was necessary to maintain normal blood sugar level. The serum insulin level by RIA was 70 IU/ml (N = 3-35 IU/ml) with a simultaneous blood sugar level of 16 mg/dl. The insulin/glucose ratio was 4.4 (N = 0.5). Serum Ca, Mg, P, Na, Cl, K, NH₃ and blood gases were within normal limits. Conservative treatment with glucagon and diazoxide at 15 mg/kg/day which was subsequently increased to 30 mg/kg/day failed to increase blood sugar to acceptable levels hence a decision to carry out a subtotal pancreatectomy was taken on day 35. After some initial fluctuations in the sugar levels and an episode of infection, the blood sugar level stabilised to above 80 mg/dl. Despite an early diagnosis and successful surgical intervention, this child is unfortunately neurologically damaged.

Pathology

No discrete nodule was palpable in the resected pancreas. Few 3 mm bits from parts of the body and tail were fixed in 3% buffered chilled glutaraldehyde, and the entire specimen blocked in paraffin and serial sections stained by H & E. Immunohistochemical stains were carried out. (courtesy Professor Julia Polak, Hamersmith Hospital, London).

Sections showed presence of small clusters of islet cells intermingled with acinar tissue scattered all over. Some of the islet cell clusters were in the centre of the acinus, suggesting an origin from centroacinar cells. Islet cell clusters could be seen budding from the larger pancreatic ducts within fibrous septa (Fig. 1). Immunostaining for neuron specific enolase sharply decorated the increased endocrine cell population. For identification of cell composition of the exocrine tissue, immunostaining for insulin, glucagon, somatostatin, pancreatic polypeptide, glicentin 1 and 2, and amylin were performed. The most striking feature was the staining for insulin, where positivity was demonstrated in numerous groups of cells throughout the pancreas (Fig. 2). Glucagon was also strongly positive but clearly lesser than insulin. The staining pattern of pancreatic polypeptide, glicentin and amylin was similar to glucagon but to a much lesser intensity. The gluteraldehyde fixed tissue was stained with uranyl acetate and lead citrate. On electronmicroscopy all four endocrine cell types A,B,D and PP were identified on basis of granule morphology. An occasional amphiocrine or composite cell was detected. Endocrine cells were demonstrated in ductules having lumen lined by microvilli, i.e., the centro-acinar cell of the terminal branch of the pancreatic duct system (Fig. 3).

Discussion

Nesidioblastosis originally described by Laidlow(3) as diffuse duct endocrine pro-
Fig. 1. Pale staining cells are seen within the acini and also budding off from the central ductule (H & E: $\times 200$).

Fig. 2. Immunoperoxidase stain for insulin reveals numerous small to medium sized clusters of beta cells intimately mixed with the exocrine tissue ($\times 80$).

liferation, is an entity associated with severe infantile hypoglycemia due to organic hyperinsulinism(4) and is occasionally seen with other endocrine tumors such as multiple endocrine neoplasia Type 2. Two basic types of lesions are associated with
Fig. 3. Ductule lined by microvilli and presence of endocrine cells in the wall (×4000).

persistent neonatal hyperinsulinemia hypoglycemia, a diffuse and a local form of nesidioblastosis. Bishop et al. (6) postulated that a substantial fall in somatostatin, which is a potent insulin inhibitor, might contribute to the development of excessive insulin release. In the present case D cells, containing somatostatin were substantially reduced. Electronmicroscopy study showed the presence of composite or amphicrine cells, a finding uniformly reported in literature. Gould et al. (7) suggested the term nesidiodyplasia to include maldistributed or malregulated endocrine and amphicrine cells, accounting for hyperinsulinism.

Infants presenting with hyperinsulinemic hypoglycemia and having morphological feature of nesidioblastosis are often cured by subtotal pancreatectomy. This is borne out by our case and similar ones reported in the literature. The practical dilemma facing the surgeon is the extent to which the pancreas must be respected. Nesidioblastosis cannot be diagnosed on frozen sections of the pancreas. In the absence of a detectable adenoma, a subtotal resection (80-85%) should be performed as in our case. This may be curative or may lead to better control with diazoxide. If desirable results are still not obtained, then a total pancreatectomy may be necessary. Wilson et al. (8) have reported that carefully monitored subcutaneous usage of somatostatin analogue (SMS 201-995) has been useful in the long term treatment of recurrent hypoglycemia after subtotal pancreatectomy.

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Paraplegia Following Cisternal Puncture in Thoracic Osteochondroma

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Solitary spinal osteochondromas are rare lesions. They grow slowly and usually manifest in the second and third decades of life. Recently we came across a case of solitary thoracic osteochondroma who developed paraplegia following a cisternal puncture for myelography. Because of its rare occurrence and rarity of this complication in bony tumor, it is being reported.

Case Report

A 14-year-old male patient was admitted to the neurological service of the hospital with complaints of difficulty in walking and weakness of both lower limbs of 20 days duration. There was no history of backache or root pains. He denied any history of trauma.

On examination, higher functions, cranial nerves and upper limbs were normal. In the lower limbs, tone was increased. Power was Grade III at the hips and Grade IV in all other groups of muscles. Deep tendon jerks were exaggerated. Ankle clonus was present, plantar were bilaterally extensor and abdominals were absent. No deficit was found in the upper thoracocervical region. Hemogram, ESR and urinalysis were normal. Plain X-ray showed scoliosis of thoracic spine and mottled calcification on the left side at D2 level. Lumbar myelogram revealed an extradural block at D3 level (Fig.) while cervical myelogram one week later through lateral cervical approach revealed the extradural block at D2 level. Following lateral cervical myelography, he developed complete loss of power in both lower limbs and graded sensory loss up to D4. D1-3 laminectomy revealed a bony mass arising from D2

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