The term small round-cell tumour (SRCT) is a generic term for tumours composed of malignant round cells that are slightly larger or double the size of red blood cells in air-dried smears or measure less than 10 μm in diameter in alcohol-fixed smears, and have scanty cytoplasm. Their features, including cellularity, morphology, pattern of cell arrangement and smear background, often pose a diagnostic challenge in FNAC since similar features may reflect a variety of tumour types and subtypes. Within the group of tumours that express a dominant or occasional SRCT (excluding the central nervous system neoplasms) include Ewing’s sarcoma and primitive neuroectodermal tumour (PNET; ES/PNET) and its variants, neuroblastoma, desmoplastic SRCT, rhabdomyosarcoma (RMS) (alveolar, solid and embryonal), small-cell osteosarcoma, chondrosarcoma (myxoid and mesenchymal), round-cell and myxoid liposarcoma, synovial sarcoma (monophasic undifferentiated), primitive malignant peripheral nerve sheath tumour (malignant small-cell schwannoma), Non Hodgkin lymphome (NHL), Merkel cell tumour of the skin and small-cell carcinoma including neuroendocrine carcinoma [1].

7.1 Small Round Cell Tumours of Childhood

Tumours of infancy and childhood are most commonly referred to as the SRCT group and include ES/PNET, neuroblastoma, RMS and malignant lymphoma. Other malignancies that may be considered in the differential diagnosis include small-cell osteogenic sarcoma, undifferentiated (anaplastic) hepatoblastoma, granulocytic sarco-
ma, blastemal-type Wilms’ tumour, and desmoplastic small-cell tumour of the peritoneum [2]. Although challenging, FNAC of childhood SRCT can be diagnostic in the majority of cases, allowing specific therapy to be given to patients with unresectable SRCT without a tissue biopsy as well as documenting recurrent and/or metastatic disease [2]. SRCTs usually have characteristic cytomorphology. However, when these tumours are undifferentiated, morphological criteria may not be sufficient to arrive at a correct diagnosis. A variety of ancillary studies including electron microscopy, immunohistochemistry, DNA ploidy, cytogenetics and FISH may provide valuable additional information for the precise characterisation of these neoplasms. Some ancillary studies may also be used for assigning these cases to prognostically significant subgroups, helping to define the most suitable chemotherapeutic regimens. Since most of these special studies require only a small amount of cellular material, FNAC is ideally suited for obtaining samples for these procedures [3].

7.1.1 Ewing’s Sarcoma/Primitive Neuroectodermal Tumour

ES/PNET is a family of malignant SRCTs that exhibits neuroepithelial differentiation, most often presenting as a soft-tissue or bone lesion in the trunk or axial skeleton, predominantly in older children and adolescents. Isolated cases of PNET have been observed in FNAC samples from visceral sites such as the ovary, testis, uterus, bladder, pancreas and kidney [4]. In some cases the primary diagnosis made by FNAC enables the paediatric oncologist to give specific therapy for the otherwise unresectable tumour and thus achieve remission [5]. Local recurrences may include the chest wall, pleura and pericardium, whilst metastatic disease may be found almost anywhere.

The cytological features of ES/PNET include malignant cells with a high N/C ratio, hyperchromatic nuclei without prominent nucleoli, distinctively smooth nuclear membrane contour, finely granular chromatin, one or two small nucleoli and scant, but almost always present, perinuclear clear cytoplasm, suggesting epithelial differentiation (Fig. 7.1). Cells are arranged singly or in cohesive clusters. Homer-Wright rosettes may be seen and there are no ganglion cells or neuropil. There is an absence of frequent mitotic figures, large nucleoli, nuclear pleomorphism, cellular debris, histiocytes and polymorphonuclear leukocytes [6]. Smears appear clean, with small, uniform cells having features suggesting a neuroendocrine epithelial tumour (Fig. 7.2). As regards the differentiation of Ewing’s sarcoma, a few subtle differentiating features can be observed: the cells in Ewing’s sarcoma have a finer nuclear chromatin in comparison to those of PNET tumour, and punched-out clear cytoplasmic vacuoles are present. PNET shows nuclear moulding, unipolar cytoplasmic tags and Homer-Wright rosettes [7].

Fig. 7.1