Animal mitochondrial DNA (mtDNA) is commonly described as a small, circular molecule that is conserved in size, gene content, and organization. Data collected in the last decade have challenged this view by revealing considerable diversity in animal mitochondrial (mt-) genome organization. Much of this diversity has been found in four phyla of nonbilaterian animals: Cnidaria, Ctenophora, Placozoa, and Porifera, which – from a phylogenetic perspective – form the main branches of the animal tree along with Bilateria. Non-bilaterian mt-genomes vary substantially in size and rate of sequence evolution, often have introns, encode a variable number of tRNAs, and utilize at least 7 different genetic codes. Furthermore, some of these genomes display linear and/or multichromosomal organization, contain extra protein genes, and their expression involves tRNA and mRNA editing as well as translational frameshifting. Here I will describe some of these unusual observations and discuss their implications for understanding and treatment of human mitochondrial diseases.